

SOLITARY AND MULTIPLE ENCHONDROMAS (OLLIER DISEASE): RARE SKELETAL DISORDERS CAUSING SEVERE BONE DESTRUCTION AND PSYCHO-EMOTIONAL DISTRESS

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SOLITARY AND MULTIPLE ENCHONDROMAS (OLLIER DISEASE): RARE SKELETAL DISORDERS CAUSING SEVERE BONE DESTRUCTION AND PSYCHO-EMOTIONAL DISTRESS (Abstract): Solitary enchondromas represent benign cartilaginous neoplasms originating from residual chondrocytes of the physal plate. Ollier disease (multiple enchondromatosis) is a rare, non-hereditary skeletal disorder characterized by multiple intramedullary cartilaginous tumors (enchondromas) carry a risk of malignant transformation into chondrosarcoma or atypical cartilaginous tumor (ACT). Both diseases are rare, initially asymptomatic and curative treatment is non-existent. **Materials and methods:** The study included a cohort of 14 patients diagnosed with solitary or multiple asymptomatic enchondromas, classified according to the Takigawa classification. In all cases the diagnosis was established by imaging examination. All patients underwent surgical treatment. The surgical techniques employed consisted of tumor curettage followed by reconstruction of the resulting bone defect using either autologous bone graft or solid bone substitute. In one case, where the enchondromas were multiple and large, with massive digital bone destruction, a necessary amputation was performed. **Results:** Imaging examinations revealed extensive lytic lesions affecting the metacarpals and phalanges in all cases. In the patient with Ollier disease, lesions were also identified in the radius, humerus, scapula, and vomer bone-an exceptional localization. Histopathological analysis confirmed a central atypical cartilaginous tumor (ACT) arising secondarily from an enchondroma in the case of multiple enchondromatosis. The patient with Ollier disease was also diagnosed with psychiatric comorbidities, including anxiety and depression, related to chronic hospitalization and physical deformity. Functional and aesthetic outcomes were evaluated in all cases using the DASH, ROM, and VAS scores. **Conclusions:** The presence of enchondromas is rare, and multiple enchondromatosis-particularly when associated with cranial involvement-represents an extremely uncommon entity. The absence of symptoms often leads to incidental diagnosis. In Ollier disease, onset occurs in early childhood, and the resulting deformities can cause significant psychological and emotional trauma, in addition to the orthopedic and functional burden associated with the disease. **Keywords:** SOLITARY ENCHONDROMA; OLLIER DISEASE; CHONDROSARCOMA; VOMER BONE ENCHONDROMA; SURGERY.

INTRODUCTION

Solitary enchondromas represent benign cartilaginous neoplasms originating from

residual chondrocytes of the physal plate. More than half of the reported cases are localized within the bones of the hand (1).

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The true incidence of this pathology is likely underestimated, as enchondromas frequently remain clinically silent. Diagnosis is most often established incidentally during routine radiographic evaluation or in response to clinical manifestations such as localized pain, swelling, or a pathological fracture. Radiographic examination typically demonstrates a well-circumscribed lytic lesion that generally preserves cortical integrity and does not involve adjacent soft tissues, with variable degrees of intralesional calcification (2, 3).

The management of benign osseous tumors of the hand, including enchondromas, typically involves intralesional curettage, with or without subsequent filling of the residual cavity using autologous bone graft or bone substitute material (4). Although autologous bone grafting remains the gold standard for reconstructing bone defects, the use of bone substitutes has become increasingly prevalent in order to minimize donor-site morbidity and associated complications. In cases requiring cancellous bone grafting for hand defects, the distal radius and the olecranon of the ulna are the most commonly utilized donor sites. These locations generally provide sufficient cancellous bone for reconstruction of defects in the carpal bones, metacarpals, and phalanges, and are preferred due to their accessibility within the same operative field and compatibility with the same regional anesthesia (5, 6).

Multiple enchondromatosis also known as Ollier disease, is a rare, non-hereditary skeletal disorder that occurs sporadically and was first described in 1889 by Louis Léopold Ollier (1, 2). It is typically diagnosed in early childhood, manifesting through painful bone deformities, limb asymmetry, and growth disturbances. The estimated incidence of the disease is approximately one case per 100,000 individu-

als, making it an exceptionally uncommon condition (3). In Ollier disease, multiple enchondromas are irregularly distributed and may appear either unilaterally or symmetrically, varying in both number and stage of development (4, 5). The diagnosis is primarily based on clinical evaluation and imaging studies, with histopathological assessment being essential to identify malignant transformation—most frequently into chondrosarcoma or, less commonly, osteosarcoma (1). The reported risk of malignant transformation ranges between 5% and 50%. Since 2013, the World Health Organization (WHO) has reclassified grade I chondrosarcoma as an atypical cartilaginous tumor (ACT), recognizing it as a locally aggressive neoplasm with limited metastatic potential and classifying it as an intermediate-grade rather than a fully malignant tumor (2, 3).

These lesions can lead to progressive bone destruction, skeletal deformities, and functional disability, often beginning in childhood and persisting into adulthood (6). The visible deformities, coupled with chronic pain and restricted movement, frequently result in psychological distress, including anxiety, low self-esteem, and depressive symptoms, which significantly affect the patient's quality of life (1, 7).

Despite being a rare and debilitating condition with a significant risk of malignant transformation, there is currently no specific curative treatment for Ollier disease (2). Patients presenting with small, asymptomatic enchondromas that do not compromise limb function are typically managed through careful clinical and radiological observation. Surgical treatment is indicated only in cases complicated by limb shortening, severe deformities, pathological fractures, or suspected malignancy (8). Depending on the extent and severity of

bone destruction, surgical approaches may include tumor excision followed by bone grafting, either using autologous bone or synthetic bone substitutes (9). The bone substitute can be solid or injectable, and may also be employed in the absence of an autologous graft (9). When cortical thinning remains significant after tumor ablation, osteosynthesis materials such as Kirschner wires, plates and screws, or external fixators can be used to reinforce bone stability and prevent postoperative fracture. In extensive lesions with severe cortical destruction where reconstruction is not feasible, amputation of the affected segment may be the only viable treatment option (10).

Advanced imaging modalities, including MRI and CT, are critical in determining the extent of the lesions, assessing cortical involvement, and detecting early malignant transformation. Bone scintigraphy may also be used to evaluate metabolic activity and identify lesions with aggressive potential (11, 12). Imaging studies are further valuable in identifying associated systemic abnormalities, as Ollier disease has been linked to conditions such as skull base gliomas, pancreatic neoplasms, and hematologic disorders, supporting the concept of a broader systemic predisposition related to somatic mosaicism (13).

We report a retrospective study including 13 patients diagnosed with unilateral solitary enchondroma of the hand and one patient diagnosed with multiple enchondromatosis (Ollier disease), with lesions also involving the vomer bone and significant psychiatric impairment.

MATERIALS AND METHODS

A total of 14 patients, aged between 23 and 54 years, were included in this study.

All were admitted to the Plastic Surgery Clinic of Sf. Spiridon County Clinical Emergency Hospital Iasi between 2016 and 2023.

The study was approved by the institutional ethics committee, and informed consent was obtained from all participants. All patients presented with slow-growing, painless deformities of the metacarpal or phalangeal bones. None of the cases reported pain or sensory disturbances. In five cases, patients presented to the emergency department with pathological fractures resulting from minor trauma, such as direct blows or falls from standing height. One patient was known to have Ollier disease (multiple enchondromatosis), diagnosed at the age of three, associated with psychiatric pathology secondary to physical deformities. The reason for medical presentation in this case was the excessive tumor volume involving both hands, particularly the second finger of the right hand. Radiological imaging was performed in all cases, while computed tomography (CT) was additionally conducted in the patient with Ollier disease. Surgical treatment was indicated in all patients. Surgical interventions were carried out under either WALANT (Wide Awake Local Anesthesia No Tourniquet) or loco-regional infraclavicular block anesthesia. All excised specimens were submitted for histopathological examination. Postoperative outcomes were assessed over a two-year follow-up period, using the DASH, ROM, and VAS scoring systems.

RESULTS

The study included 14 patients aged between 23 and 54 years (mean age 38.5 years), of whom 12 were male (85.7%) and 2 were female (14.3%). Thirteen patients presented with unilateral solitary enchondroma of the hand, while one patient was

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diagnosed with multiple enchondromatosis (Ollier disease), representing 7.1% of the cohort (tab. I).

TABLE I.

Baseline Characteristics

Variable	Value
Total patients	14
Age range (years)	23-54
Mean age (approx.)	38.5
Sex: Male	12 (85.7%)
Sex: Female	2 (14.3%)
Lesion type: Solitary	13 (92.9%)
Lesion type: Multiple (Ollier)	1 (7.1%)
Laterality	Unilateral in 13, bilateral in 1 (Ollier)
Anatomic sites (hand)	11 phalanges, 1 metacarpal, 1 multifocal (Ollier)

Among the solitary cases, enchondromas were localized in 11 patients at the level of the phalanges—specifically, two cases involved first phalanx of middle finger of the left hand, five cases first phalanx of the fifth finger of the right hand, three cases first phalanx of the fifth finger of the left hand, and one case third phalanx of the fifth finger of the left hand. One lesion was situated in the second metacarpal.

The patient with Ollier disease presented with multiple lesions affecting both hands (phalanges and metacarpals), bilateral radius, humerus, scapula, and, exceptionally, the vomer bone (fig. 1).

Radiological exam confirmed the presence of well-circumscribed lytic lesions in all cases (fig. 2).



Fig. 1. Clinical aspect of the both hands. a. dorsal aspect, b. palmar aspect

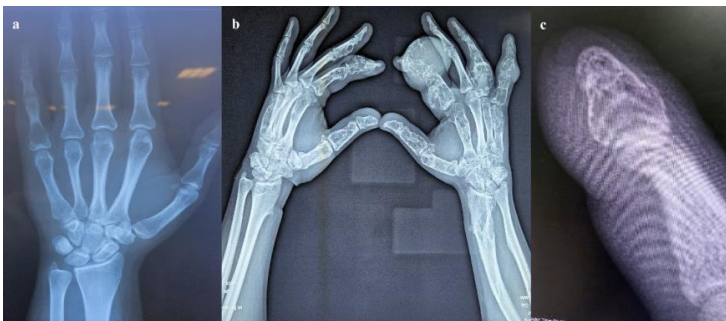


Fig. 2. Radiological aspect of a solitary enchondroma a. first phalanx of the fifth finger, b. distal phalanx of the medius finger, c. multiple enchondromatosis

The lesions typically demonstrated cortical thinning without rupture, endosteal scalloping, and occasional intralesional calcifications. No periosteal reaction or soft-tissue extension was observed. These radiographic features were consistent with the benign appearance of enchondromas and supported the indication for conservative surgical curettage.

For the patient with Ollier disease, combined clinical and imaging assessments established the diagnosis of multiple enchondromatosis (Ollier disease), which was subsequently confirmed by genetic testing. Multiple surgical interventions had been performed throughout childhood and adolescence. By the age of 18, the patient's medical history included Ollier disease, a pathological spiral fracture of the distal third of the left femur with displacement (surgically treated in 2010), left genu valgum (surgically corrected in 2024), limb length discrepancy with a 9 cm shortening of the right lower leg, bilateral knee stiffness, and generalized growth delay. Given the cumulative burden of prolonged hospitalizations, repeated surgeries, skeletal deformities and asymmetries, combined with a disrupted family and social environment, the patient was diagnosed with an organic personality disorder, chronic sleep disturbances, generalized anxiety disorder, and severe depressive episodes with anxiety paroxysms. He has been under psychiatric care since the age of 16. Between the ages of 18 and 26, the patient did not seek medical follow-up regarding the progression of his condition. At the time of current admission, clinical examination revealed marked deformities involving the metacarpals and phalanges of both hands, which were asymmetrical and varied in size.

Preserved and symmetrical cutaneous

sensitivity was noted. In the right hand, both active and passive opposition movements were impaired due to limited flexion of the 2nd, 4th, and 5th digits, resulting in an inability to perform a cylindrical grasp. Additionally, all previously documented skeletal deformities were clinically confirmed.

The CT scan reveals, in case of multiple enchondromatosis, multiple expansile lytic bone lesions consistent with enchondromas, some exhibiting heterogeneous structure with internal calcifications. Several lesions demonstrate cortical disruption with soft tissue extension and significant bone deformities, particularly in the hand skeleton. No periosteal reactions are noted. The lesions are more numerous in the right upper limb. In the right radius, at the distal third, multiple lytic expansile lesions with cortical breach and mild soft tissue extension are observed, with a maximum size of 12×25 mm.

In the right humerus, a central, expansile lytic lesion with calcifications is seen in the proximal metaphysis, causing cortical thinning without periosteal reaction, measuring 20×34×36 mm. A similar centro-medullary lesion with calcifications (9×13 mm) is noted in the mid-diaphysis, with mild asymmetric cortical thinning. The right scapula (partially included in the scan) shows multiple lytic lesions, particularly in the coracoid process, with the largest measuring 18 mm. The left humerus has a large, central osteolytic lesion extending from the proximal metaphysis into the diaphysis, occupying nearly half its length, with cortical thinning and focal erosions, particularly in the metaphyseal region, with a cranio-caudal diameter of 110 mm. The left scapula (partially included) contains similar lytic lesions, mainly in the coracoid

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process, with the largest measuring 14 mm. Additional lesions are seen in the pisiform and triquetral bones, with significant cortical lysis in the pisiform. In the left hand, numerous lytic lesions affect the phalanges, albeit smaller than those on the right. Some exhibit cortical thinning, focal disruptions, and extra-osseous extension, with the large-

est at the first phalanx of the thumb measuring 16×13×17 mm. Small osteolytic lesions are also present in the carpal bones, some with cortical thinning. In the left radius, a central osteolytic lesion in the proximal metaphysis shows mild expansion and cortical thinning, measuring 13 mm (fig. 3).

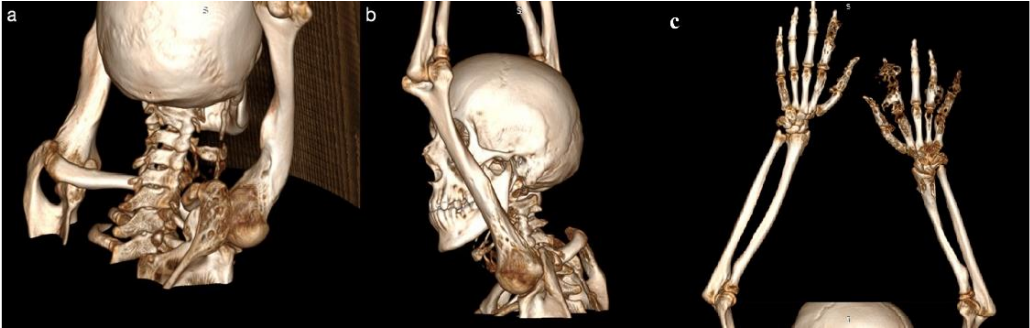


Fig. 3. CT imaging. a, b. Enchondromas located in bilateral humerus and the left scapula.

In the right hand, extensive lytic lesions affect the metacarpal bones and phalanges, sparing only the third digit. Most lesions show cortical thinning or focal disruptions, with a particularly large, expansile lesion in the second digit (notably involving the proximal and middle phalanges), measuring 48×50×39 mm, with gross cortical des-

truction and extra-osseous extension, c. Enchondromas involving the metacarpals and phalanges of both hands, as well as the distal end of the left radius

Additional findings include an expansive lytic lesion with cortical irregularities and interruptions in the vomer bone, associated with nasal septum deviation to the left (fig. 4).

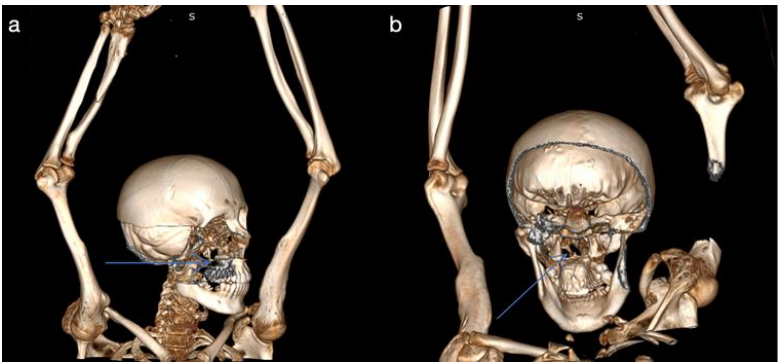


Fig. 4. CT appearance of an enchondroma located in the vomer bone- destruction of the vomer bone: a. lateral view, b. posterior view.

A C2-C3 vertebral block is present, and, despite significant artifacts, cystic areas are noted in the posterior fossa (19×23 mm), suggestive of arachnoid cysts or a megacisterna magna. The right thyroid lobe is heterogeneous, containing a hypodense nodule of 9×8 mm, and a similar posterior left thyroid nodule of 9×8 mm is also noted.

Based on the patient's history, clinical findings, and imaging results, surgical intervention was deemed necessary at the level of the second digit of the right hand, which was causing significant functional impairment. This limitation represented the main reason the patient sought medical attention (tab. II).

TABLE II.
Operative Details

Parameter	Findings
Anesthesia	8 loco-regional (infraclavicular block), 6 WALANT (1: 100,000 dilution)
Type of surgical procedure	Intralesional curettage ± cavity filling in 13 cases; amputation in 1 case (Ollier)
Osteosynthesis material	Used in 13/14 cases (92.9%)
Bone grafting	Autologous bone graft in 3 cases (21.4%)
Bone substitute	Solid bone substitute in 10 cases (71.4%)
Fixation stability	Achieved in all reconstructed cases
Fixation stability	Achieved in all reconstructed cases
Amputation (necessity)	1 case - Ollier disease, large tumor (6.5×5×5.8 cm)

Loco-regional anesthesia (infraclavicular block) was used in eight patients (57.1%), while the WALANT technique (1: 100,000 dilution) was used in six (42.9%). Surgical management consisted of intralesional curettage with or without cavity

filling in 13 cases (92.9%), and amputation was required in one case due to massive bone destruction secondary to multiple enchondromatosis.

For reconstruction, autologous bone grafts were used in three patients (21.4%), whereas solid bone substitutes were applied in ten (71.4%). Osteosynthesis material was employed in 13 of 14 cases (92.9%) to ensure structural stability (figs. 5, 6).

In the patient with multiple enchondromatosis, amputation was required due to the considerable tumor dimensions (6.5×5×5.8 cm) and the presence of extensive osseous destruction.

All curetted specimens were submitted for histopathological analysis. In patients with solitary enchondromas, no signs of malignant transformation were identified (0/13) (tab. III).

In contrast, histopathological evaluation of the giant tumor in the patient with multiple enchondromatosis revealed a mesenchymal tumor proliferation involving the phalangeal structures of the second digit of the right hand, characterized by a multilobular growth pattern, consistent with a secondary atypical cartilaginous tumor (ACT). The lesion was composed of an abundant cartilaginous matrix, forming multiple lobules of irregular size and shape, separated by fibrous septa with congested vasculature. Areas of both low and high cellularity were observed. The chondrocytes showed minimal to moderate cytonuclear pleomorphism, with well-preserved eosinophilic cytoplasm and irregular, unequal, hyperchromatic nuclei. The nuclei displayed variable morphology, ranging from lymphocyte-like to stellate and fusiform shapes.

Occasional typical mitotic figures were identified. Focally, regions with marked pleo-morphism and increased density of atypical tumor cells were also noted (fig. 9).

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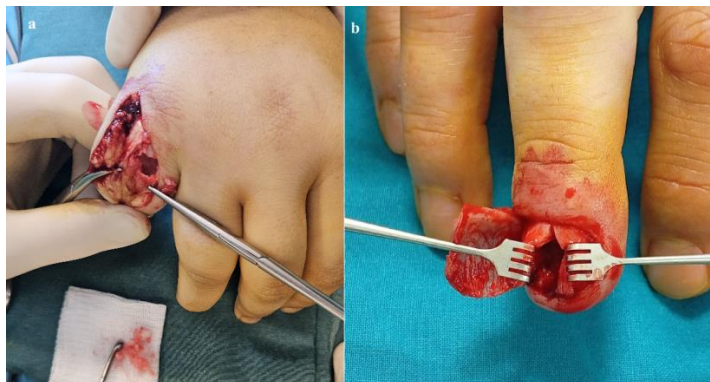


Fig. 5. Some cases of solitary enchondroma - intraoperative view:
a. first phalanx of the fifth finger, b. distal phalanx of the middle finger



Fig. 6. Postoperative radiological aspect - bone reconstruction with solid bone substitute:
a. First phalanx of the fifth finger, b. Distal phalanx of the middle finger

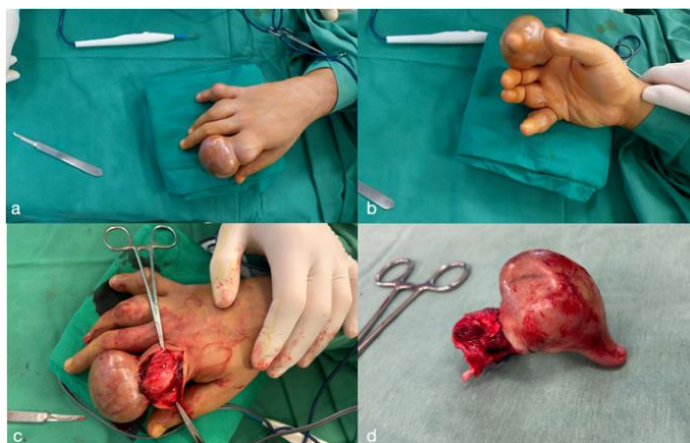


Fig. 7. Preoperative dorsal view: a - dorsal aspect, b.- palmar aspect,
c. intraoperative view, d. amputated segment (the entire right index finger)

TABLE III.
Imaging and Histopathological Findings

Variable	Findings
Imaging findings	Lytic lesions in all 14 cases (100%)
Solitary lesions	No malignant transformation identified (13/13)
Multiple enchondromatosis (Ollier)	Mesenchymal tumor proliferation with multilobular growth pattern, consistent with secondary ACT

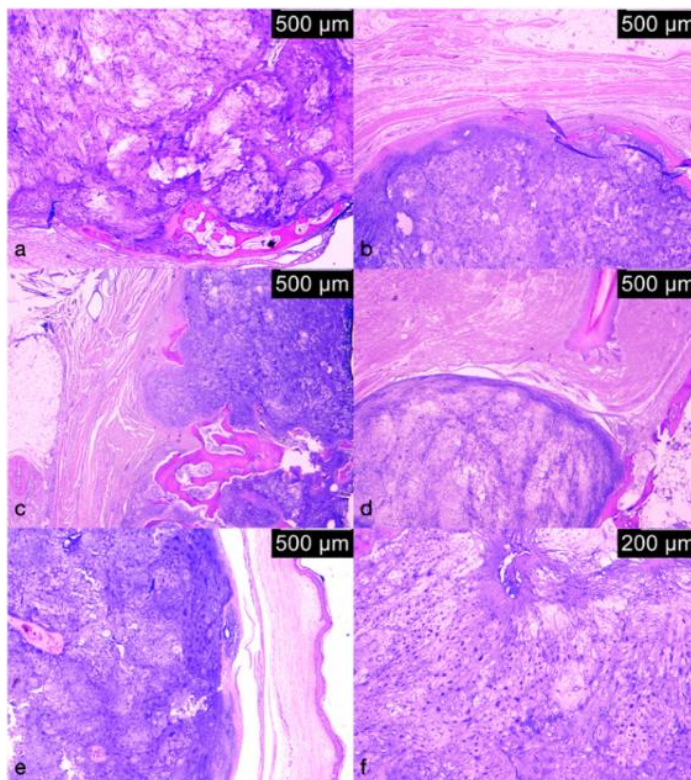


Fig. 8. a. ACT-cortical bone destruction with invasion into adjacent connective tissue ($\times 2.5$); b. ACT- cortical bone destruction with invasion into an adjacent ligament ($\times 2.5$); c. ACT- cortical destruction with soft tissue invasion ($\times 2.5$); d. ACT- invasive nodule in subcutaneous tissue ($\times 2.5$); e. ACT-evidence of invasion at the lobular front ($\times 2.5$); f. ACT-chondrocytes with moderate cytonuclear atypia ($\times 5$).

All patients demonstrated favorable postoperative evolution, with no local recurrence or infection. At three months, complete wound healing and stable osteosynthesis were noted in all cases.

By six months, radiographic evaluation

confirmed structural integrity and bone consolidation, with preserved joint mobility and satisfactory aesthetic appearance (tab. IV).

The mean DASH score at final follow-up was 8.4 ± 3.2 , indicating minimal disa-

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bility. The range of motion (ROM) recovery ranged between 80-100% compared to the contralateral side, and the mean VAS pain score at six months was 1.2/10. Patient satisfaction, both functional and aesthetic, was uniformly high. In the patient diagnosed with Ollier disease, psy-

chiatric therapy was maintained throughout hospitalization under the supervision of the consulting specialist. The patient was discharged with a recommendation for reevaluation after three months; however, follow-up was not achieved due to financial constraints.

TABLE IV.
Outcomes and Follow-up

Outcome parameter	Findings
Follow-up duration	2 years
3-month outcomes	Favorable in all cases; wound healing complete; no infection or recurrence
6-month outcomes	Stable bone structure, no recurrence; good joint mobility
DASH score (mean \pm SD)	8.4 \pm 3.2 (indicative of minimal disability)
ROM recovery	80-100% compared to contralateral side
VAS pain score at 6 months	Mean 1.2/10
Patient satisfaction (subjective)	High in all cases (no aesthetic complaints)
Psychiatric follow-up (Ollier case)	Maintained during hospitalization; follow-up lost due to financial constraints

DISCUSSION

Solitary enchondroma represents the most common benign cartilaginous tumor of the hand, typically arising from residual chondrocytes of the physal plate. Although histologically benign, its clinical behavior and radiological appearance may mimic low-grade chondrosarcoma, particularly in cases with cortical expansion or endosteal scalloping. The true incidence of enchondroma is likely underestimated, as many lesions remain asymptomatic and are discovered incidentally during imaging performed for unrelated reasons. In the present series, 13 of 14 patients (92.9%) were diagnosed with solitary enchondroma, most frequently involving the phalanges—an observation consistent with previous reports indicating a predilection for the small tubular bones of the hand (1-3). The phalanges, particularly the proximal and middle segments of the ulnar digits, are

known sites of increased enchondral ossification activity, which may explain the predominance of these locations.

Clinically, most patients presented with slowly progressive, painless swelling or localized deformity, in agreement with findings from Kuur and associates (4), who emphasized the insidious nature of the lesion. In this cohort, pain was not a presenting symptom in any case, and functional limitation was minimal prior to surgical intervention. In five cases, however, patients presented to the emergency department with pathological fractures following minor trauma—a typical manifestation of enchondroma-related bone fragility. Radiographically, all lesions appeared as well-defined lytic areas with cortical preservation and variable intralesional calcifications, consistent with the classical imaging features described in the literature (5). Advanced imaging such as CT or MRI is

generally reserved for cases where the extent of cortical involvement or suspicion of malignancy must be clarified. Surgical curettage remains the standard of care for solitary enchondromas of the hand, as it effectively removes the lesion while preserving the structural and functional integrity of the digit. In the present study, all patients underwent intralesional curettage, followed by reconstruction of the residual bone cavity using either autologous cancellous bone graft or a solid bone substitute. Autologous bone grafts were preferred in younger patients with smaller defects, while substitutes were used in larger lesions to minimize donor-site morbidity. These findings align with the observations of Gaulke *et al.* (6), who reported comparable consolidation times and recurrence rates between autologous grafts and synthetic substitutes. Osteosynthesis was required in nearly all cases to restore mechanical stability after curettage. No post-operative complications or recurrences were recorded during the two-year follow-up period, confirming the efficacy of this surgical approach. The functional outcomes, as measured by DASH and ROM scores, were excellent across the cohort, with a mean DASH score of 8.4 and pain scores approaching zero at six months.

Overall, the outcomes reported in this series reinforce the current understanding that solitary enchondromas of the hand, when managed by meticulous curettage and appropriate reconstruction, have a benign course, excellent functional recovery, and negligible risk of recurrence or malignant transformation. An association between Ollier disease and mutations in the parathyroid hormone receptor 1 (PTHRI) gene, was proposed, particularly the R150C variant, which alters signaling

pathways regulating chondrocyte proliferation. Subsequently, in 2011, Amary *et al.* identified somatic mosaic mutations in the IDH1 and IDH2 genes—encoding isocitrate dehydrogenase enzymes - as central to the disease mechanism (5, 14). These mutations result in aberrant production of D-2-hydroxyglutarate, an oncometabolite that interferes with normal epigenetic regulation, leading to disorganized endochondral ossification, abnormal chondrocyte differentiation, and increased tumorigenic potential (5, 15). Clinically, Ollier disease presents with pain, swelling, and visible deformities, most frequently affecting the hands and lower limbs, often accompanied by limb-length discrepancies resulting from asymmetric skeletal growth. The condition typically progresses slowly; however, the cumulative effects of multiple enchondromas may lead to severe functional impairment, skeletal deformity, and aesthetic disfigurement, profoundly impacting daily activity and psychological well-being (16, 17). Pathological fractures are common, particularly in long bones weakened by extensive enchondromatous replacement (18). The bilateral distribution of lesions in the hands, as seen in this case, is particularly unusual, since Ollier disease typically exhibits unilateral or asymmetrical involvement. Bilateral disease likely reflects a broader embryonic distribution of the somatic mutation, suggesting that the genetic alteration occurred earlier during embryogenesis, giving rise to a wider population of affected mesenchymal progenitor cells. This mechanism may explain the multifocal and systemic pattern observed in this patient, involving not only the hands but also the radius, humerus, scapula, and notably the vomer bone (19, 20). In the present case, the

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enchondromatous masses were large, producing extensive bone destruction, with CT imaging revealing near-complete resorption of the phalanges. The disease course is chronic and unpredictable, marked by alternating periods of relative stability and episodes of accelerated lesion growth that may herald incipient malignant transformation. In the reported case, histopathological examination of the amputated segment revealed malignant changes within the enchondroma, consistent with transformation into an atypical cartilaginous tumor (ACT).

The craniofacial localization of an enchondroma in the vomer bone represents an exceptional and clinically significant finding. Enchondromatous lesions in cranial bones are exceedingly rare, with only a handful of cases reported in the literature. Their presence challenges the conventional understanding of the anatomical and embryological boundaries of endochondral ossification, suggesting a wider reach of the somatic mosaicism underlying the disease. The involvement of a flat skull-base bone, such as the vomer, supports the hypothesis that Ollier disease can manifest in nontraditional skeletal regions derived from endochondral ossification centers, including the sphenoid, occipital, and vomer bones. Craniofacial involvement may further complicate the clinical course by producing neurological or otorhinolaryngological manifestations, such as nasal obstruction, sinonasal expansion, or, in rare cases, intracranial extension. In this patient, however, the vomer lesion was entirely asymptomatic, discovered incidentally on CT imaging without associated nasal or sinus manifestations. This rare presentation broadens the current understanding of Ollier disease, reinforcing its multisystemic

and mosaic nature and emphasizing the importance of comprehensive imaging evaluation in detecting atypical skeletal localizations that might otherwise remain clinically silent (21, 22).

The surgical management of Ollier disease remains challenging and somewhat controversial. While curettage and bone grafting may be effective for localized lesions, extensive or multifocal disease significantly limits reconstructive potential. Amputation, although radical, may be justified in cases of massive deformity, functional loss, or histologically confirmed malignant transformation. In the present case, amputation of the affected digit provided rapid functional improvement, with no postoperative complications, underscoring the importance of individualized, function-oriented management rather than strict anatomical preservation (23).

Beyond orthopedic considerations, Ollier disease exerts a profound psychological impact. The combination of visible deformities, chronic pain, and recurrent hospitalizations leads to marked emotional distress, particularly in patients diagnosed during childhood (24, 25). The current case illustrates the cumulative psychosocial burden associated with the disease: early-life trauma, together with persistent physical disfigurement and repeated medical interventions, contributed to the development of chronic depression, anxiety, and organic personality disorder. Similar reports in the literature describe the emotional strain of chronic deformity and disability, but few documents psychiatric morbidity of this intensity (26-28).

The literature describes cases of multifocal disease with cranial extension, yet none report such severe psychiatric comorbidity, highlighting the unique multidimen-

sional complexity of the present case—where skeletal deformity, oncologic risk, and psychological decline intersect in a single, rare clinical scenario.

CONCLUSIONS

Ollier disease imposes a profound functional and psychosocial burden on affected individuals. Progressive skeletal deformities of the hands result in substantial loss of dexterity, compromised grasping ability, and a marked decline in overall quality of life. However, the psychological consequences of the disorder often surpass its physical manifestations. Chronic disfigurement, prolonged hospitalization, and

persistent social stigmatization contribute to severe emotional distress, manifesting as anxiety, depressive episodes, and long-term psychological exhaustion.

These findings underscore the necessity of a multidisciplinary approach that integrates surgical management with continuous psychological and social support, aimed at optimizing both functional outcomes and mental well-being.

CONFLICT OF INTERESTS AND FUNDING

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