

Primary Costal Bone Xanthoma

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ABSTRACT

Primary bone xanthoma is an exceptionally infrequent clinical entity. It predominantly affects young men, with a higher occurrence in flat bones of the axial skeleton. This pathology can present in an isolated (primary) form or as secondary, associated with endocrinological and metabolic disorders such as hyperlipidemia. It is always necessary to investigate and rule out the secondary cause to certify normolipidemia. The therapeutic approach should be tailored to the lesion characteristics, location, and neighboring structures' affectation.

We present the case of a female patient with no significant history who had an incidental finding of a costal lytic lesion on computed tomography. Due to the risk of fracture and the inability to rule out malignancy, we performed a resection of the body of the fifth right rib, guided by video-assisted thoracoscopic surgery. The histopathological study reported a medullary cavity with infiltration of foamy macrophages and numerous cholesterol crystals, confirming the diagnosis of bone xanthoma.

Key words: bone xanthoma, histiocytes, hyperlipidemia, normolipidemia, video-assisted thoracoscopy.

Xantoma óseo costal primaria

RESUMEN

El xantoma óseo primario es una entidad clínica extremadamente infrecuente. Afecta en mayor proporción a hombres jóvenes, con mayor presentación en huesos planos del esqueleto axial. Esta patología puede aparecer en forma aislada (primaria) o secundaria (asociada a trastornos endocrinológicos y metabólicos como la hiperlipidemia). La causa secundaria siempre debe investigarse y descartarse, certificando normolipidemia. El abordaje terapéutico del xantoma debe adecuarse a las características de la lesión, su ubicación y la afectación de las estructuras vecinas.

Presentamos el caso de una paciente de sexo femenino sin antecedentes de relevancia, con hallazgo incidental de lesión lítica costal por tomografía computarizada. Debido al riesgo de fractura e imposibilidad de descartar malignidad se realizó resección de cuerpo de quinta costilla derecha guiado por videotoracoscopia. El estudio histopatológico informó: cavidad medular con infiltrado de macrófagos espumosos y numerosos cristales de colesterol, confirmando el diagnóstico de xantoma óseo.

Palabras clave: xantoma óseo, histiocitos, hiperlipidemia, normolipidemia, toracoscopia videoasistida.

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CLINICAL CASE

A 57-year-old female patient with a history of arterial hypertension and normolipidemia (total cholesterol 200 mg/dL, triglycerides 167 mg/dL) attended a consultation with the thoracic surgery team after being referred due to an incidental finding of a rib lesion on a chest computed tomography scan performed in the context of trauma. The lesion was reported as a lytic expansile lesion measuring 44 × 12 mm on the fifth right costal arch without invasion of muscular structures (Fig. 1). On questioning she denied pain, discomfort, ventilatory mechanics alterations, or any associated symptoms. Physical examination revealed no palpable mass. We performed a total body bone scan to rule out the presence of other associated lesions, which showed a solitary lesion (Fig. 2). Additionally, we did a CT-guided percutaneous biopsy; the histopathological study reported numerous macrophages, multinucleated giant cells, deposits of hemosiderin, and cholesterol crystals, with no evidence of cellular atypia, consistent with a probable bone xanthoma (BX). Initially, given the characteristics of the lesion, differential diagnoses

included enchondroma, osteblastoma, osteosarcoma, and Erdheim-Chester disease.

Given that it was a lytic lesion with a risk of fracture in a young patient and malignancy could not be categorically ruled out, surgical resection was decided. We performed a resection of the body of the right fifth rib under video-assisted thoracoscopy with 3 cm margins and sent the specimen to Pathology for deferred study. The patient had an uncomplicated postoperative course and was discharged on the fifth day with good pain management.

In the pathology of the lesion, histological sections showed a medullary cavity with infiltration of foamy macrophages and numerous cholesterol crystals. Multinucleated giant cells, areas of fibrosis, and a few lymphocytes and plasma cells were also observed (Fig. 3), leading to a diagnosis of costal bone xanthoma (BX).

DISCUSSION

Bone xanthoma (BX) is an uncommon pathology that usually presents in young male patients. These lesions are frequently observed in superficial soft tissues, although

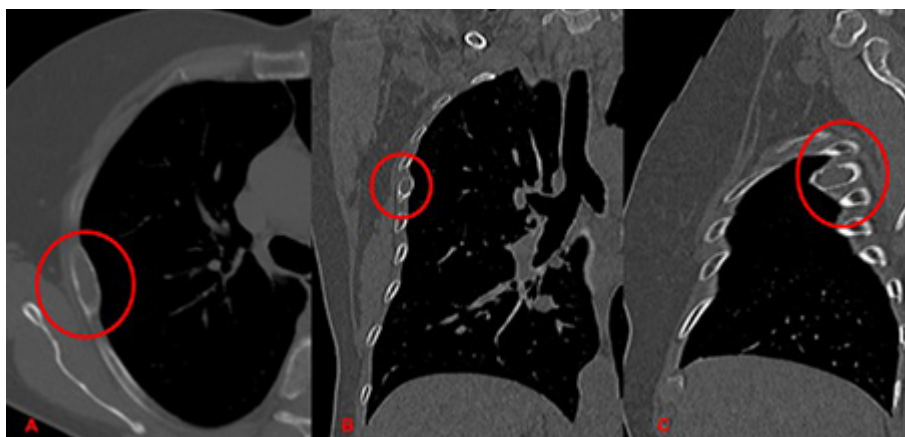


Figure 1. Computed tomography showing a lytic lesion in the midsection of the fifth right costal arch. A. Axial view. B. Coronal view. C. Sagittal view.



Figure 2. Scintigraphy with increased tracer uptake in the lesion of the 5th right costal arch.

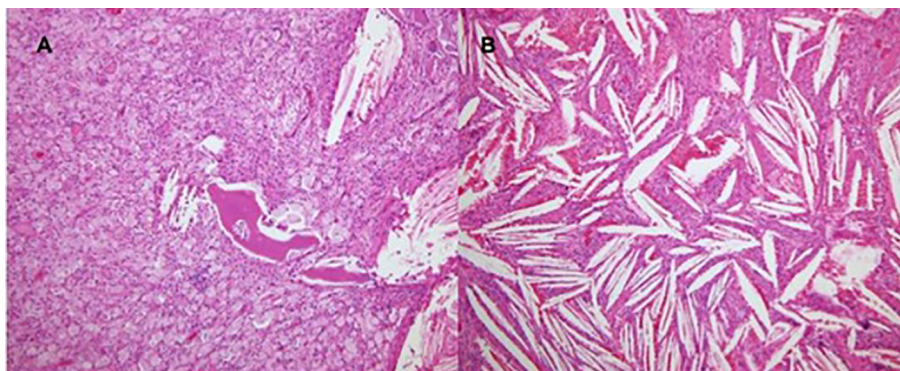


Figure 3. A. Hematoxylin-eosin 10×: medullary spaces occupied by clusters of foamy macrophages and cholesterol crystals are observed. B. Hematoxylin-eosin 40×: foamy macrophages at higher magnification.

there are scarce reports published of their appearance in deep structures such as bones.¹⁻⁶ The most common anatomical sites reported in the literature include pelvic bones, skull, ribs, humerus, and cervical vertebrae, with a greater predisposition to affect flat bones.¹ Bertoni et al. reported that, out of 21 cases, only 3 involved the rib cage. In the reviewed literature, the ages of presentation ranged from 7 to 60 years,^{1,7} with a male-to-female ratio of 2:1, making reports of female patients like ours rare.

These lesions are often secondary to systemic metabolic diseases such as hyperlipidemia, endocrinological or systemic conditions like Erdheim-Chester disease, Langerhans cell histiocytosis, or post-traumatic fibrous dysplasia.⁵ There are also cases associated with trauma and traumatic fractures. Another less common form of presentation is primary, where these lesions are associated with normal serum cholesterol levels and without other history related to the pathology, as in our case, where we ruled out secondary causes that could provoke it.

Regarding the clinical presentation of this pathology, most patients may experience pain or compressive symptoms related to the location of the lesion; however, it is also less frequently found as an incidental finding in asymptomatic patients, as in our case. The average duration of symptoms ranges from 2 weeks to 5 years, with an average of 17.8 months.¹ The imaging diagnosis is often complex and complicated to interpret. Several reports show that BX can present on computed tomography studies as isodense images in T1 sequences and hyperdense in T2 sequences,^{7,8} There are also reports where it frequently appears as osteolytic lesions⁶, as in the case presented in this text. In the presence of a suspected bone xanthoma (BX) in imaging studies, we recommend bone scintigraphy to assess multifocal lesions and for subsequent follow-up.¹

Treatment for this pathology can range from conservative management to radical surgeries. In the case of medical treatment for patients with dyslipidemia, reducing serum cholesterol levels through lifestyle changes, a low-fat diet, and the use of lipid-lowering drugs such as statins and chenodeoxycholic acid is described,

along with biannual follow-up with radiological controls.⁹ Surgical options vary depending on the lesion's location, size, and relationship with neighboring structures. Once malignancy is ruled out, curettage and bone grafting are indicated in the pelvis or tibia. In other locations, such as the spine and skull base bones, adjuvant therapies like radiation have been used.⁴ For rib lesions, especially those smaller than 5 cm that do not involve multiple ribs, soft tissues, lung, or pleura, various surgical options exist, always maintaining margins of 2-3 cm.¹⁰ In our case, since the lesion was neither visible nor palpable, we chose a combined approach, guided by video-assisted thoracoscopy for identification and marking of the lesion, followed by resection through a mini-thoracotomy. That is one of the few cases requiring rib cage resection for definitive treatment.¹¹

CONCLUSION

Bone xanthoma is a rare benign condition. As it is impossible to reach a definitive diagnosis based on imaging alone, it usually has a surgical indication, and it is the pathological anatomy alone to confirm the diagnosis. Always investigate for an underlying secondary cause. For xanthoma, the therapeutic approach should adapt to the involvement of neighboring structures.

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Conflicts of Interest: The authors declare no conflicts of interest.

REFERENCES

1. Bertoni F, Unni KK, McLeod RA, et al. Xanthoma of bone. *Am J Clin Pathol.* 1988;90(4):377-384. <https://doi.org/10.1093/ajcp/90.4.377>.
2. Marqués Mateo M, Puche Torres M, Miragall Alba L, et al. Primary mandibular bone xanthoma. A case report. *Int J Oral Maxillofac Surg.* 2004;33(8):806-807. <https://doi.org/10.1016/j.ijom.2003.12.006>.

3. Ali S, Fedenko A, Syed AB, et al. Bilateral primary xanthoma of the humeri with pathologic fractures: a case report. *World J Radiol.* 2013;5(9):345-348. <https://doi.org/10.4329/wjrv.v5.i9.345>.
4. Alden KJ, McCarthy EF, Weber KL. Xanthoma of bone: a report of three cases and review of the literature. *Iowa Orthop J.* 2008;28:58-64.
5. Ortega Hrescak MC, Jerez JM, Arévalo CM y cols. Xantoma intraóseo pelviano: reporte de un caso y revisión de la literatura. *Rev Argent Radiol.* 2014;78(3):161-167. <https://doi.org/10.1016/j.rard.2014.07.005>.
6. Huang CF, Cheng SN, Hung CH, et al. Xanthoma of bone in a normolipidemic child: report of one case. *Acta Paediatr Taiwan.* 2000;41(3):158-160.
7. Asano K, Sato J, Matsuda N, et al. A rare case of primary bone xanthoma of the clivus. *Brain Tumor Pathol.* 2012;29(2):123-128. <https://doi.org/10.1007/s10014-011-0073-x>.
8. Yokoyama E, Ito J, Tokiguchi S, et al. [A case of xanthoma of the skull]. *Rinsho Hoshasen.* 1990;35(9):1057-1060.
9. Ahmed G, Al Dosari M, El-Mahi M, et al. Primary xanthoma of calcaneus bone: case report. *Int J Surg Case Rep.* 2014;5(10):699-702. <https://doi.org/10.1016/j.ijscr.2014.07.016>.
10. Ocakcioglu I, Sayir F. Rib resection using a Gigli saw under thoracoscopic guidance. *Wideochir Inne Tech Maloinwazyjne.* 2019;14(1):126-132. <https://doi.org/10.5114/wiitm.2018.77266>.
11. Mottola E, Adotti F, Pernazza A, et al. Xanthoma of rib: a case report and review of the literature. <https://doi.org/10.21203/rs.3.rs-1561164/v1>.