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CASE REPORT

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Primary xanthofibroma in the calcaneus: a case report

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Xanthoma or xanthofibroma is a lesion, characterized by foamy histiocytes (xanthoma cell) and is mostly seen in soft tissue. Xanthoma may also occur in in the skeletal system of patients with an abnormal lipid metabolism. We present a 22-year-old man with primary xanthofibroma in the calcaneus, who was treated by curettage and grafting of the lesion.

Key words: Calcaneus; xanthofibroma; xanthoma.

Xanthoma (or xanthofibroma) is a benign lesion, most commonly seen in the form of nodules in the soft tissue of hyperlipidemic patients. Xanthoma, developing secondary to hyperlipidemia, may also be seen in the skeletal system. The nature of this lesion is controversial. Some authors consider this condition to be a remission or degeneration of a primary underlying disease or lesion, rather than a real tumor.[1-3] Other authors, however, accept it as a real tumor.[4-7] We present a calcaneal fibroxathoma in a young patient with normal lipid metabolism and discuss our findings in combination with the data available in the literature.

Case report

A 22-year-old male presented with heel pain which had started seven months previously, without any trauma. He had no systemic disease. In physical examination, he showed tenderness to palpation of the calcaneus. Ankle movements were within the normal range. There was no increased temperature around the heel or any mass that could be felt. The neurological examination results were normal. All laboratory tests were within normal limits. Lateral radiographs of the foot showed a cystic lesion in the calcaneus with a well-defined, sclerotic border (Fig. 1). No intralesional calcification was seen. In the CT scan a lesion with a sclerotic border and cortical expansion and destruction was detected (Fig. 2). In the T1-weighted MR scan the lesion appeared hypointense and in the T2-weighted MR scan it appeared hyperintense (Figs. 3a and b). As the clinical and radiological examinations were inconclusive, an incisional biopsy was performed. The histopathological examination suggested the diagnosis of xanthoma. Consequently, the patient was treated by curettage and autogenous bone grafting. Macroscopic examination of the specimen revealed it to be a soft mass, light-brown and yellowish in color (Fig. 4). The surrounding bone tissue was sclerotic.

Varying densities of multinucleated giant cells, clusters of focal lymphocytes and connective tissue cells of fusiform character were observed amongst a large number of histiocytes with a foamy cytoplasm (Fig. 5). These pathological findings led to a diagnosis of xanthoma. There were no findings to suggest

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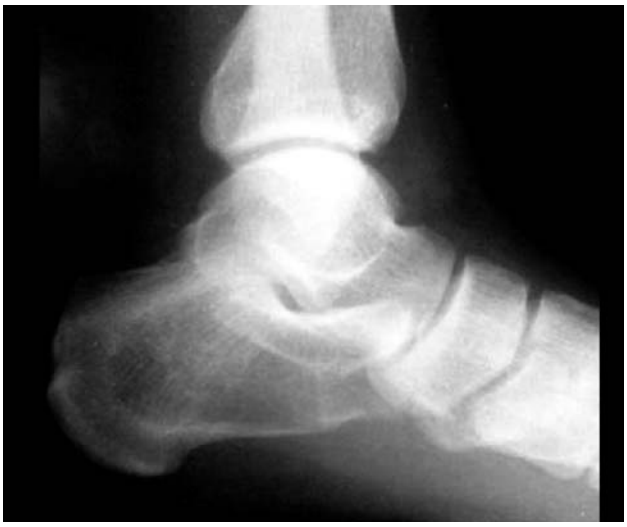

Fig. 1. Lateral radiograph of the foot. Note the lesion with a well-defined and thin sclerotic margin.

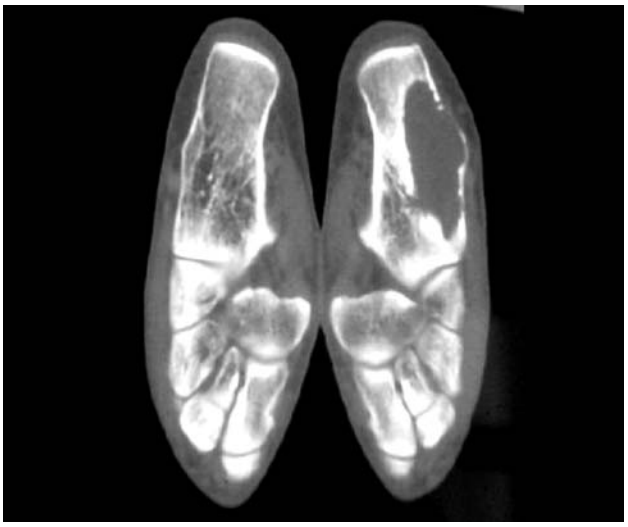

Fig. 2. CT scan showing a lytic lesion causing cortical expansion and destruction of the lateral cortex of the calcaneus.

any bone tumor. The cultures of the excised material remained sterile. There was no recurrence or heel pain by the postoperative 15th month.

Discussion

Xanthoma is a benign proliferative lesion, mostly seen in tissue vulnerable to trauma, such as the skin, subcutaneous tissue and tendon sheath of hyperlipidemic patients. Histologically, it is a lesion characterized by lipid-loaded histiocytes (foamy histiocytes, xanthoma cells), giant cells, granulation tissue and inflammatory cells. Although xanthoma is usually known as a soft tissue lesion, it may also be localized in the skeletal system. The cellular composition of xanthoma lesions localized in the skeletalsystem is quite variable, and may differ from case to case.[4] The fact that this tumor is rarely seen in the skeletal system, and the variability in cellular com position of xanthoma localized in the skeletal system causes disagreements about whether this lesion is a real tumor. Some authors define this lesion as a variant, because cells, such as xanthoma and giant cells, which have histological features similar to xanthoma, may be seen in many non-neoplastic and neoplastic lesions of bone.[1,2] For example, xanthomatous changes have been reported to occur in lesions, such as fibrous dysplasia, giant cell tumor of the bone, aneurysmal bone cysts, non-ossified fibroma, fibrous cortical defect, benign and malignant fibrous histiocytoma, Erdheim-Chester disease, xanthogranulomatous osteomyelitis, and metastatic renal cell carcinoma.[4,8-11] For this reason, it has been suggested that xanthomas should be considered as a variant of the primary lesion, rather than a real tumor. Furthermore, it has been suggested that accepting xanthomas as separate tumors would have no benefit, and that it would be more appropriate to classify them as a sub-group of benign fibrous histiositoma.[1] Xanthoma, however, may develop not only as a condition secondary to hyperlipidemia in the skeletal system of patients, but may also occur as a primary condition.[4,6,7,12-16] It is difficult to explain primary and secondary lesions with the "variant theory". In fact, it is impossible to

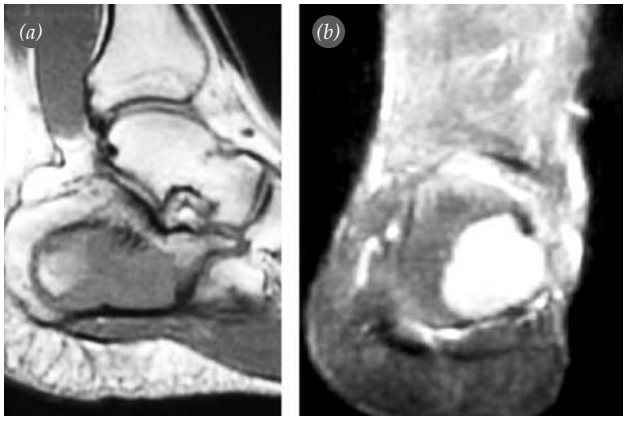

Fig. 3. In **(a)** the T1-weighted MR scan a hypointense lesion, and in **(b)** the T2-weighted MR scan a hyperintense lesion is seen.

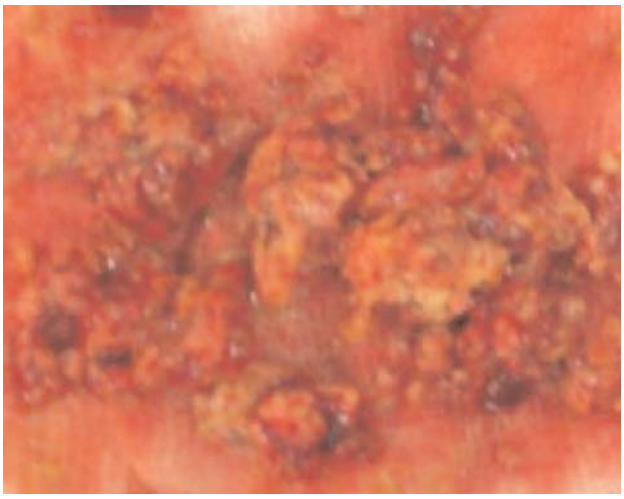

Fig. 4. Macroscopic image of the lesion. [Color figure can be viewed in the online issue, which is available at www.aott.org.tr]

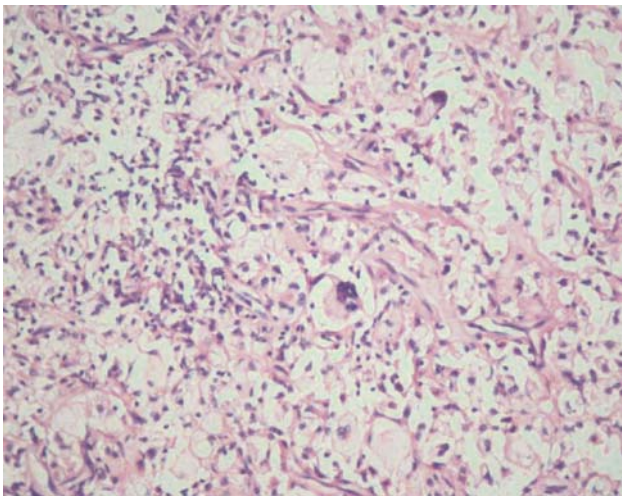

Fig. 5. Histiocytes with foamy cytoplasm, giant cells, lymphocytes and connective tissue cells (H-E x20). [Color figure can be viewed in the online issue, which is available at www.aott.org.tr]

reach a conclusion from a single case. However, when the available data in the literature is also taken into account, we think that treating all xanthomas seen in the skeletal system as belonging to the same group, or defining them all as a "variant" can bring about important errors. In this respect, we feel that it would be more correct to classify xanthomatous lesions of the skeletal system under three groups, as studying these lesions in different groups may help to eliminate the confusion in the terminology, and also allow a systematic diagnosis and treatment. The first group of xanthomas are advanced stage benign and malignant tumors of the skeletal system with xanthomatous changes.[4,8,9] These types of findings should be taken as evidence of the degeneration or remission of the primary lesion, and therefore, it may be appropriate to define these types of xanthomatous changes as "xanthomatous variant" or "variant xanthofibroma". The second type of xanthoma is one that generally forms in the skeletal system of Type-2 and Type-3 hyperlipidemic patients.[3,9,13,15] It may be appropriate to call this type of lesion a "secondary xanthoma", as it forms secondary to the hyperlipidemia and heals upon successful treatment of hyperlipidemia, without further surgical intervention. A third type of xanthoma is one that occurs as a primary disease, without any underlying cause, in any part of the skeletal system.[4,6,7,14,16] Patients with this type of xanthoma have normal lipid metabolism. Also there are no underlying non-neoplastic or neoplastic lesions. We think that it is appropriate to term this type of xanthoma "primary xanthoma". Primary xanthoma has rarely been reported in the skeletal system. Those that have been reported are mostly localized in flat bones, such as the mandible, temporal bones, ribs and pelvis. Amongst these reports, there are three cases involving the calcaneus (Table 1).[4,7,17] Primary xanthoma is more fre-quently seen in men. Generally

Table 1. Calcaneal xanthofibroma cases presented in literature.

Case	Age and sex	Complaint	Size	X-ray	Treatment	Recurrence
Bertoni et al.[4]	Unreported	-	-	Cystic lesion, sclerotic border	-	No
Yamamoto et al.[7]	51, female	Pain	4x3	Cystic lesion, sclerotic border, irregular trabeculae	Curettage + Tricalcium Phosphate	No
Kinberg[17]	19, female	No pain, coincidental	3x3	Cystic lesion, sclerotic border	Curettage + Allograft + Bone marrow	No
Kapukaya et al.	22, male	Pain	3x3	Cystic lesion, sclerotic border	Curettage + Autograft	No

it is seen in patients over 20 years of age. The most frequent symptom with xanthoma is pain. A small number, however, are discovered coincidentally. Plain radiographs present a well-defined, lytic lesion, surrounded by a sclerotic border. However, some lesions may progress with an aggressive cortical expansion and destruction, as in malignant lesions.[6,7,13,15,18,19] Intralesional calcification may be noticed in some cases. In our case, plain radiographs revealed cortical expansion, as well as cortical destruction and a well-defined partially sclerotic border. CT and MR findings were similar to those in the previous articles. These findings required a differential diagnosis to eliminate the possibility of tumors, such as solitary bone cysts, intraosseous lipoma and osteoblastoma, because both bone cysts and intraosseous lipoma are tumors which are relatively frequently localized in the calcaneus. Cholesterol accumulation may also be seen in some bone cysts due to bleeding. In our case, there was no macroscopic or microscopic cyst formation. Furthermore, histological examination of the lesion revealed no lipomatous component or osteoblastic activation.

This lesion can be quite difficult to diagnose, because it is rarely localized in the skeletal system. Imaging methods reveal no findings that are specific to the lesion, and the image it presents is mostly atypical. Differential diagnosis should be taken seriously because xanthomatous findings may be seen in the histological results of even malignant tumors of the skeletal system. Although some authors feel it is unimportant to differentiate between different types of xanthomas, we do not share this view. From our point of view, a variant xanthoma is not an original xanthoma, but one in which the histological image of the primary tumor has changed. Thus, to avoid an erroneous diagnosis, all material obtained from these lesions should be microscopically examined. Moreover, the radiological features of the primary lesion should be studied in more detail using imaging methods because we need to bear in mind that there are very important tumors amongst the lesions causing variant xanthoma. Furthermore, it is necessary to differentially diagnose between primary xanthoma and secondary xanthoma. In some cases, the first indication of hyperlipidemia may be a lesion in the skeletal system.[9] Therefore, after all diagnoses of xanthomatous lesions, the lipid metabolism of the patient should also be investigated. It should be noted that the treatment for secondary xanthoma is not surgical, and that the skeletal manifestations will disappear with systemic treatment of hyperlipidemia.[13] On the other hand, despite being a secondary xanthoma, if it has reached a size likely to cause fractures or if a pathological fracture has occurred, then surgical treatment with curettage, grafting and internal fixation should be considered.[13] Primary xanthoma may be treated with partial or total curettage, followed by an autograft. We are of the opinion that total curettage is generally more appropriate because the diagnosis of variant xanthoma can usually be made following the histological examination of the whole of the material from the lesion.

Primary xanthoma is a lesion rarely seen in the skeletal system and it is difficult to diagnose using only clinical and imaging techniques. To avoid erroneous diagnosis in the histopathological evaluation, a detailed microscopic examination should be carried out on all material obtained from these lesions and primary xanthofibroma should be considered when making the diagnosis.

Conflicts of Interest: No conflicts declared.

References

1. Macdonald D, Fornasier V, Holtby R. Benign fibrohistiocytoma (xanthomatous variant) of the acromion. Arch Pathol Lab Med 2002;126:599-601.
2. Fechner RE, Mills SE. Tumors of bones and joints. In: Rosai J, editor. Atlas of tumor pathology. Third series, fascicle 8. AFIP, Washington DC: Armed Forces Institute of Pathology; 1993. p. 161-3.
3. Huang GS, Huang CW, Lee CH, Taylor JA, Lin CG, Chen CY. Xanthoma of the sacrum. Skeletal Radiol 2004;33: 674-8.
4. Bertoni F, Unni K, McLeod RA, Sim FH. Xanthoma of bone. Am J Clin Pathol 1988;90:377-84
5. Yalçınkaya U, Öztop F. Xanthoma of the calcaneus associated with hyperlipoproteinemia. J Am Podiatr Med Assoc 2005;95:602-4.
6. Kuroiwa T, Ohta T, Tsutsumi A. Xanthoma of the temporal bone. Case report. Neurosurgery 2000;46:996-8.
7. Yamamoto T, Kawamoto T, Marui T, Akisue T, Hitora T, Nagira K, et al. Multimodality imaging features of primary xanthoma of the calcaneus. Skeletal Radiol 2003;32:367-70.
8. Vankalakunti M, Saikia UN, Mathew M, Kang M. Xanthogranulomatous osteomyelitis of ulna mimicking neoplasm. World J Surg Oncol 2007;5:46.
9. Dallari D, Marinelli A, Pellacani A, Valeriani L, Lesi C, Bertoni F, et al. Xanthoma of bone: first sign of hyperlipi-

demia type IIB: a case report. Clin Orthop Relat Res 2003;(410):274-7.

10. Blanco M, Cabello-Inchausti B, Cura M, Fernandes L. Post-traumatic fibro-osseous lesion of the ribs and scapula (sclerosing xanthofibroma). Ann Diagn Pathol 2001;5: 343-9.
11. Dalinka MK, Turner ML, Thompson JJ, Lee RE. Lipid granulomatosis of the ribs: focal Erdheim-Chester disease. Radiology 1982;142:297-9.
12. Khandpur S, Manchanda Y, Sharma VK, Singh MK, Tandon N. Rare association of xanthoma disseminatum with skeletal involvement. J Dermatol 2003;443:190-3.
13. Yokoyama K, Shinohara N, Wada K. Osseous xanthomatosis and a pathologic fracture in a patient with hyperlipidemia. A case report. Clin Orthop Relat Res 1988;(236): 307-10.
14. Friedman O, Hockstein N, Willcox TO Jr, Keane WM. Xanthoma of the temporal bone: a unique case of this rare condition. Ear Nose Throat J 2000;79:433-6.
15. Inserra S, Einhorn TA, Vigorita VJ, Smith AG. Intraosseous xanthoma associated with hyperlipoproteinemia. A case report. Clin Orthop Relat Res 1984;(187):218-22.
16. Mateo MM, Torres PM, Miragall Alba L, Iglesias Gimilio ME, Pascual Gil JV. Primary mandibular bone xanthoma. A case report. Int J Oral Maxillofac Surg 2004;33:806-7.
17. Kinberg P. Xanthoma of a calcaneus. J Foot Ankle Surg 1998;37:531-4
18. Inserra S, Einhorn TA, Vigorita VJ, Smith AG. Intraosseous xanthoma associated with hyperlipoproteinemia. A case report. Clin Orthop Relat Res 1984;(187):218-22.
19. Fink IJ, Lee MA, Gregg RE. Radiographic and CT appearance of intraosseous xanthoma mimicking a malignant lesion. Br J Radiol 1985;58:262-4.