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INTRAMUSCULAR MYXOMA AND FIBROUS DYSPLASIA OF BONE – MAZABRAUD'S SYNDROME

A case report

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Abstract

We present a case of Mazabraud's syndrome, a rare benign disease, with multiple intramuscular myxomas of the thoracic wall associated with fibrous dysplasia of bone. CT, MR imaging and ultrasonography (US) of the thorax showed 2 well circumscribed homogeneous intramuscular tumors. A US-guided needle biopsy with a large-core needle (2.0 mm) and a fine needle (0.8 mm) showed that the tumors were intramuscular myxomas with no sign of malignancy. ^{99m}Tc bone scintigraphy showed a markedly increased uptake in the right lower skull, and multiple smaller foci. CT of the skull revealed a right-sided unilateral bone thickening of the orbit and the ethmoidal cells, and right-sided exophthalmia. This case history suggests that patients with multiple intramuscular myxomas should be preoperatively examined for osseous lesions. A postoperative follow-up should also be performed to detect other soft-tissue myxomas not as yet clinically detectable, or rare osseous complications.

Key words: Intramuscular myxoma, fibrous dysplasia of bone; soft-tissue tumors; ultrasound-guided biopsy.

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Mazabraud's syndrome is a rare benign disease, characterized by the association of intramuscular myxoma and fibrous dysplasia of bone. We present a case with multiple myxomas of the thorax and affection of the skull. The preoperative diagnosis was established by an ultrasound-guided needle biopsy of the soft-tissue tumor of the thoracic wall. Fibrous dysplasia of bone was discovered during the diagnostic work-up, and was predominantly located in the skull.

Clinical history

A 50-year-old woman noticed a discrete lesion on the dorsal side of the right hemithorax. Thirty years previously she had a benign tumor excised from the right orbital edge. The nature of this tumor was unknown. Also, 10 years previously, an intramuscular myxoma had been removed from the right lateral thoracic wall.

CT, MR imaging and ultrasonography (US) of the thoracic wall showed a well circumscribed, homogeneous, lobulated intramuscular tumor, measuring 6×4×2 cm. CT showed a hypodense, nonenhanced tumor. MR showed 2 intramuscular tumors, thus revealing a smaller tumor more medially situated. Both tumors were hypointense on T1-weighted sequences, with an inhomogeneous increase in signal intensity after contrast medium injection, and hyperintense on T2-weighted images with fat suppression. US showed a solid, homogeneous, markedly hypoechoic tumor (Fig. 1). A color Doppler investigation showed no intratumoral flow. A US-guided needle biopsy with a large-core needle (Tru Cut 2.0 mm) and a fine needle for aspiration (0.8 mm) was performed. Microscopy showed an intramuscular myxoma, with no sign of malignancy. ^{99m}Tc bone scintigraphy showed a markedly increased uptake in the right lower skull, and multiple smaller foci in the ribs, sternum and spine (Fig. 2). CT of the skull

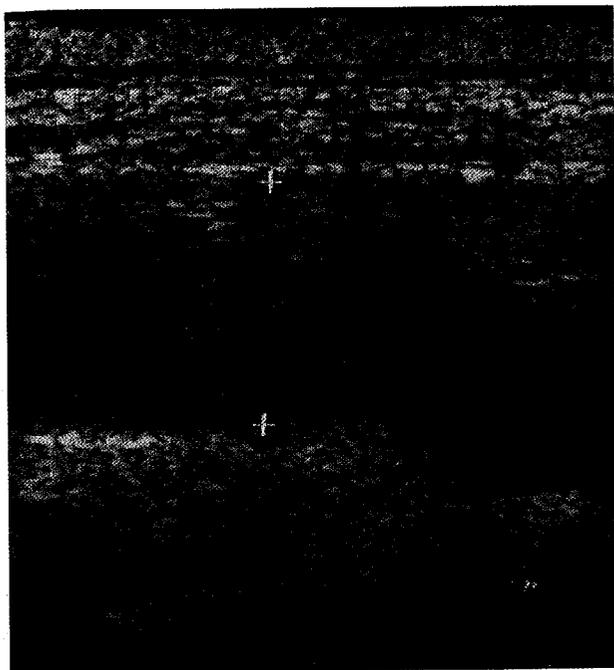


Fig. 1. US of the thoracic wall shows a homogeneous, markedly echopoor, lobulated intramuscular tumor.



Fig. 2. ^{99m}Tc bone scintigraph shows an increased uptake in the right upper face, sternum and several ribs.

showed unilateral diffuse bone thickening of the right orbit and ethmoidal cells, causing a right-sided exophthalmia, but no sign of ocular tumoral involvement (Fig. 3).

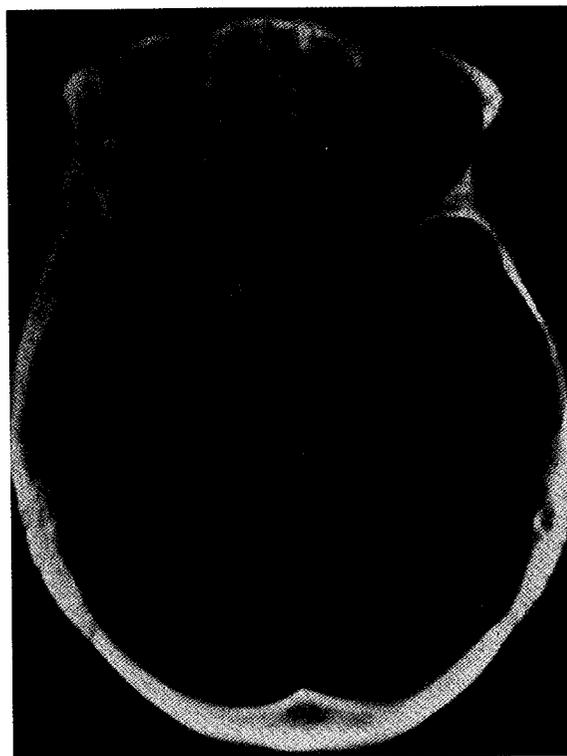


Fig. 3. CT of the skull shows right unilateral bone thickening of the orbit and ethmoidal cells.

At surgery, 2 encapsulated intramuscular tumors were found and excised *in toto*. The pathological examination confirmed the diagnosis, and showed an abundant mucoïd material with a small number of spindle-shaped and stellate cells with no pleomorphism. Small cystic spaces and scattered vessels were seen.

Discussion

Intramuscular myxoma is an uncommon benign soft-tissue tumor, characterized by the presence of an abundant myxoid matrix similar to Wharton's jelly, the material seen in the core of the umbilical cord of the mature fetus (8). Most intramuscular myxomas are solitary and occur in the 6th and 7th decades of life (8, 12). They are often located in the large muscles of the thigh, shoulder and buttocks (6, 8). CT and MR examinations are essential in the preoperative planning of excision of soft-tissue tumors. The CT findings in our patient were consistent with previous descriptions (1, 2). The MR appearance was in agreement with previously reported cases (1, 4, 6, 13).

High-resolution US is not commonly used in the evaluation of soft-tissue tumors. It can, however, show a characteristic pattern in intramuscular

myxomas, revealing hypoechoic tumors, with small fluid-filled clefts and cystic areas, as reported of 3 out of 4 patients in a recent review (3). This was not seen in our patient, who had a homogeneous, markedly hypoechoic tumor. We noticed an absence of intratumoral flow at color Doppler examination, in accordance with a previous report (3). Pathological examination of intramuscular myxomas usually shows a circumscribed, often nonencapsulated, tumor with interdigitations with surrounding muscle fibers. This might suggest that the tumor is locally aggressive but reports tend to show that recurrences are exceptional (8, 12).

Fibrous dysplasia of bone is a rare benign condition of unknown origin, with fibro-osseous metaplasia of one (monostotic) or multiple (polyostotic) bones. The long bones, ribs and skull are the sites most commonly affected (6). Complications can occur, such as fractures or – rarely – malignant transformation (5, 11).

HENSCHEN in 1926 presented a case of osteitis fibrosa associated with multiple myxomas (7). Similar cases were then sporadically reported (9, 10). MAZABRAUD et al. (11) reviewed the literature in 1967 and proposed the existence of a syndrome, characterized by the association of intramuscular myxoma (single or multiple) and fibrous dysplasia of bone. A recent review of the literature brought the total of registered cases to 26 (1). The syndrome is equally frequent in women and men, and is seen at all ages.

The present case is typical: the patient had multiple myxomas (6) and multiple osseous lesions (5, 15). Intramuscular myxomas tend to be located in the lower extremities (1, 15) and in close proximity to bones most severely affected by fibrous dysplasia (1, 5, 8, 15), although this was not the case in our patient. Unlike most reported cases (1, 15) the osseous lesions in our patient had not been discovered at the time this patient's thoracic tumors were detected. It was the ^{99m}Tc bone scintigraphy that revealed the important bone lesions in the orbital region, so attention was oriented towards the patient's face and her exophthalmia. We do not have the pathological diagnosis of the tumor excised 30 years previously from the right orbital edge, but it may well have been the patient's first intramuscular myxoma.

The etiology of Mazabraud's syndrome is unknown. The affliction is believed to be caused by a basic metabolic error of both soft and bone tissues during the initial growth period (15). The role of a somatic gene mutation has also been discussed (1, 9). An association with the McCune-Albright syndrome has been described but our pa-

tient had no cutaneous lesions or endocrinal disturbances.

In a patient with multiple and sometimes large intramuscular tumors that are associated with bone lesions, this rare syndrome should be kept in mind. An incorrect preoperative diagnosis of neurofibroma, lipoma (15) or myxoid liposarcoma (1) could thus be avoided. Differentiation from these other soft-tissue tumors can only be made by a preoperative biopsy. A correct pathological diagnosis can often be established easily and safely by a US-guided large-core needle biopsy (3), which avoids large cystic areas (14). Although the tumor is benign, postoperative follow-up should be undertaken in order to detect other myxomas not as yet clinically detectable, or rare osseous complications, i.e. malignant transformation or pathological fracture.

Conclusion: This patient had several muscular tumors of the thoracic wall and was examined by CT, MR and US. Preoperative pathological diagnosis was established by a US-guided needle biopsy. ^{99m}Tc bone scintigraphy revealed the associated bone lesions of the skull. A patient with multiple intramuscular myxomas and fibrous dysplasia of bone is at risk of developing other myxomas that are not as yet clinically detectable, or rare osseous complications.

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