Parachordoma/myoepithelioma

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Received: 16 January 2012 / Revised: 4 April 2012 / Accepted: 5 April 2012 / Published online: 20 May 2012
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Diagnosis

Parachordoma/myoepithelioma

A 44-year-old male with enlarging soft tissue mass in index finger: diagnosis and discussion

Discussion

The MRI signal characteristics of parachordomas are largely nonspecific [1]. On T1-weighted images, this particular lesion is heterogeneously isointense to muscle with peripheral elements of T1 hyperintensity, suggesting interdigitated fat. There is also signal heterogeneity with lobular areas of increased signal intensity on T1-weighted images. Mild peripheral enhancement is seen on post-contrast images. Although the initial impression on MRI of a lesion in a digit that is closely related to a tendon, in this case the extensor digitorum tendon, would be a giant cell tumor of the tendon sheath, the signal characteristics suggest otherwise. A giant cell tumor would be expected to be predominantly hypointense on both T1- and T2-weighted images due to

hemosiderin, and variably enhance with intravenous contrast [2]. The noncystic nature of the lesion excludes a ganglion cyst. The presence of interdigitated fat at the periphery of this lesion could also suggest the diagnosis of a hemangioma, but the mild increased post-contrast enhancement makes hemangioma unlikely, as hemangiomas typically avidly enhance (unless thrombosed) and often have a serpentine configuration [3].

Histologically, this lesion showed a lobular architecture with a chondro-myxoid background with some interdigitation of fat peripherally. There was minimal cellular atypia and no mitoses. The tumor cells were round to oval with intra-cytoplasmic vacuoles. The tumor was positive for AE1/AE3, CAM 5.2, EMA, S-100, vimentin, and CK 903. It was negative for CD 34 and GFAP. The proliferation marker Ki-67 was positive in 15 % of cells.

The morphology was consistent with parachordoma (myoepithelioma) [4]. Most giant cell tumors of the tendon sheath are moderately cellular and composed of sheets of rounded or polygonal cells that blend with hypocellular collagenized zones. Variable numbers of giant cells are usually present. Hemosiderin-containing xanthoma cells are also common and often localized at the periphery of giant cell tumors [5]. The morphology of our lesion was quite different.

Hemangiomas on histology would be expected to show dilated blood-filled spaces and discrete vessels [6], which is not demonstrated in this case. Therefore, the combination of MRI and histological features suggests that the most likely diagnosis is a parachordoma.

Parachordomas (or myoepitheliomas) are rare soft tissue tumors with fewer than 100 cases reported [1]. They histologically may resemble chordomas, and occur in a nonaxial

The case presentation can be found at doi:10.1007/s00256-012-1413-6.

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location. They have an indolent nature with a male predilection, typically occurring in the fourth decade of life and usually in the extremities [7]. There have been reported late recurrence and metastasis [8, 9]. Parachordoma originally was believed to be a chordoma occurring in nonaxial sites, but now parachordomas are considered a unique entity [10].

The nomenclature of these lesions is not settled. Parachordoma was a term used because of the histological resemblance to chordoma (central chordoma). The name myoepithelioma was suggested several years ago for these lesions. Some authors have described true brachyury positive peripheral chordomas that are separate from parachordoma/myoepithelioma. Recent genetic discoveries on these tumors have re-kindled this debate, since some of these have a translocation involving the EWSR1 gene. The current WHO fascicle lists the tumor as mixed tumor/myoepithelioma/parachordoma with the understanding that all three terms are appropriate until consensus is reached. Once a large series of these tumors are characterized by FISH/genetic studies, we may have a better understanding of these interesting lesions.

The management for these lesions is wide surgical excision, with clinical and imaging follow up to exclude recurrence.

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