US and MRI appearance of a collagenous fibroma (desmoplastic fibroblastoma) of the shoulder

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Abstract Collagenous fibroma (desmoplastic fibroblastoma) is a recently described rare benign tumor. Since this tumor was characterized by Evans in 1995, fewer than 100 cases have been reported in English literature with the largest series published by Miettinen and Fetsch. Preoperative radiological diagnosis is important to avoid over-treatment and unnecessary extensive procedures, but is difficult because diagnostic imaging findings for collagenous fibroma have not been established. Here we report the US and MRI appearance of a lesion localized deep under the deltoid muscle in a 34-year-old woman with a palpable painless mass. Histological findings were consistent with a collagenous fibroma. As far as we know, this is the first description of US appearance of a collagenous fibroma in English literature.

Riassunto Il fibroma collagenico (fibroblastoma desmoplastico) è un tumore benigno raro di recente descrizione. A partire dalla sua caratterizzazione da parte di Evans nel 1995, meno di 100 casi sono stati riportati nella letteratura inglese con la più larga casistica pubblicata da Miettinen e Fetsch. La diagnosi radiologica preoperatoria è importante per evitare eccessivi trattamenti e procedure non necessari, ma è difficoltosa perché non sono ancora stati definiti dei

reperi imaging diagnostici per il fibroma collagenico. Qui presentiamo l'aspetto ecografico e RM di una lesione localizzata in profondità rispetto al muscolo deltoide in una donna di 34 anni con una massa palpabile non dolente. I reperti istologici erano indicativi di un fibroma collagenico. Per quanto è a nostra conoscenza, si tratta della prima descrizione dell'aspetto ecografico di un fibroma collagenico riportata nella letteratura in lingua inglese.

Keywords Collagenous fibroma · Desmoplastic fibroblastoma · Soft tissue lesion

Introduction

Collagenous fibroma (desmoplastic fibroblastoma) is a benign fibrous soft tissue tumor first described by Evans in 1995 [1]. Since then, the largest series of this lesion, with 63 cases, was published by Miettinen and Fetsch [2] in 1998. This tumor is characterized by spindle-shaped to stellate fibroblastic cells, sparsely distributed in a dense fibrous background [3]. It shows a gradual growth, commonly without aggressive local infiltration. Neither local recurrence nor metastases have been reported after resection [4].

Diagnostic imaging findings of this lesion have not been clearly defined.

Here we report ultrasound (US) and magnetic resonance imaging (MRI) findings of a collagenous fibroma of the shoulder.

Case-report

A 34-year-old woman presented with a palpable painless mass of the anterior prospect of the left shoulder. US

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Fig. 1 Axial US scan, anterior aspect of the shoulder: the lesion lies deeply under the deltoid muscle, just anterior to the coracoid process; the deep aspect of the overlying deltoid muscle appears imprinted in this point. The mass has lobulated, rather defined margins and it appears isoechoic to the muscle with a few inhomogeneous hypoechoic spots

Fig. 2 US sagittal scan, anterior aspect of the shoulder: the power Doppler displays rich diffuse vascularity of the lesion

revealed a solid mass (long axis 33 mm), localized deeply under the deltoid muscle, anterior to the coracoid process (Fig. 1). The lesion was isoechoic to the muscle, with smooth and lobulated margins, apparently showing an expansive rather than an infiltrative behavior: the deltoid muscle appeared in this point imprinted by the most superficial lobule of the mass. Power Doppler evaluation displayed a rich diffuse vascularization of the mass (Fig. 2). MRI showed intermediate signal intensity (isointense to muscle) on T1-weighted images and rather homogeneous mild high signal intensity with sporadic scattered low signal intensity areas on STIR and T2-weighted images (Fig. 3a). Post-contrast T1-weighted images with fat suppression demonstrated inhomogeneous

enhancement with scattered areas of no enhancement (Fig. 3b).

The lesion was surgically removed and analyzed. Grossly, it appeared as a well-circumscribed mass, with a firm consistency and a gray-yellow color, with no signs of invasion to adjacent tissues. Histologically the lesion showed a fibroblastic proliferation with a rich stromal component that was prevalently loose with myxoid areas and sporadic malignant foci. S100 protein was not expressed. The pathologic diagnosis was collagenous fibroma.

Discussion

Collagenous fibroma, also known as desmoplastic fibroblastoma, is a benign fibrous soft tissue tumor first described by Evans in 1995. This tumor primarily occurs in the subcutaneous tissues or skeletal muscle in adults; few cases have been encountered in children. It has a peak incidence in the fifth to seventh decades of life with a significant male predominance [1]. The tumor has a wide anatomic distribution, but arms, shoulders and neck are relatively frequent sites [4]. Miettinen et al. [2] analyzed 63 cases of collagenous fibroma from the files of the Armed Forces Institute of Pathology and showed involvement of the arm (24 %), shoulder girdle (19 %), posterior neck or upper back (14 %), feet or ankles (14 %), leg (14 %), hand (8 %), and abdominal wall and hip (6 %). The patients typically presented with a history of a painless, slowly growing mass, often of relatively long duration. The tumors ranged in size from 1 to 20 cm (median 3.0 cm). The lesions were predominantly subcutaneous, but fascial involvement was common, and 27 % of cases involved skeletal muscle [2].

Grossly, collagenous fibroma appears as a well-circumscribed mass with a white to gray cut surface. Histologically, the lesion is hypocellular and consists of spindle-to stellate-shaped cells embedded in a dense collagenous or myxocollagenous stroma. Mitotic figures are rare or absent and necrosis is not present. Collagenous fibroma may be mistaken for several other benign and low-grade malignant soft tissue tumors, including desmoid-type fibromatosis and low-grade fibromyxoid sarcoma. Clonal chromosomal aberrations have been detected by cytogenetic analysis in 10 cases of collagenous fibroma. The long arm of chromosome 11, in particular 11q12, is involved in all cases except one [1].

Treatment of collagenous fibroma is surgical excision with no reported incidence of local recurrence or metastases [5].

Preoperative radiological diagnosis is important to avoid over-treatment and unnecessary extensive procedures, but

Fig. 3 **a** Sagittal TSE T2 fat sat MR scan: the lesion, showing lobulated but well defined margins, is located anterior to the coracoid process, predominantly in the fat tissue deeply under the deltoid muscle, which in this point appears thinned and compressed; the mass displays hypersignal with sporadic scattered low signal intensity

areas; no signal abnormalities are found in the surrounding structures; **b** axial SE T1 fat sat MDC MR scan: the lesion shows diffuse but inhomogeneous contrast enhancement; its margins are not clearly defined on the deeper aspect

is difficult because diagnostic imaging findings for collagenous fibroma have not been established [4].

Few case-reports described the imaging appearance of this lesion, especially MRI features, and heterogeneous aspects were found.

Shuto et al. [5] described the computed tomography (CT) and MRI appearance of a collagenous fibroma in the left lower abdomen. CT showed an inhomogeneous mass with an inhomogeneous enhancement. MRI demonstrated an inhomogeneous low signal intensity on T1-weighted images and mixed signal intensity (areas of low and high signal intensity) on T2-weighted images. Post-contrast T1-weighted images showed irregular, non-enhancing areas within an otherwise markedly enhanced mass.

Yamamoto et al. [4] described MR imaging findings of three collagenous fibromas. All three cases showed areas of low to slightly high signal intensity on T2-weighted images and areas of low to intermediate signal intensity on T1-weighted images. On post-contrast T1-weighted images with fat suppression a poor or mild internal enhancement was observed in all three collagenous fibromas, probably reflecting the limited vascularity. A rim enhancement was also noted in all three cases and was considered to represent the abundant vascularity of the outer capsule-like fibrous tissue in contrast to the inside of the tumor.

Kamata et al. [3] described the MRI evolution of a collagenous fibroma of the thigh. It showed inhomogeneous signal on T1-weighted images, with signal predominantly isointense with muscle, but including several areas of low signal intensity. On T2-weighted images, the mass predominantly showed low signal intensity with scattered

high signal intensity areas. T1-weighted images after contrast agent administration revealed heterogeneous enhancement of the lesion with areas of no enhancement. After 1 year no changes in pattern were detected on MRI despite increasing size.

In our case the MRI features of the lesion were intermediate signal intensity (isointense to muscle) on T1-weighted images and rather homogeneous mild high signal intensity with sporadic scattered low signal intensity areas on STIR and T2-weighted images; post-contrast T1-weighted images with fat suppression demonstrated inhomogeneous enhancement with scattered areas of no enhancement.

At our knowledge in English literature there are no descriptions of US aspect of collagenous fibroma. In our case US revealed a solid mass, isoechoic to muscle, with smooth and lobulated margins. It was localized in the fat deeply under the deltoid muscle and showed an expansive rather than an infiltrative behavior, apparently compressing the muscle itself in this point. Power Doppler evaluation displayed a rich diffuse vascularization of the mass.

The histological report demonstrated the presence of sporadic malignant foci within the lesion and this fact has never been reported in the English literature, with the exception of a case of a collagenous fibroma of the bone (distal femur) in a 41-year-old woman, described in 2010 by Hye Sook Min et al. [6].

Conflict of interest Mara Bonardi, Vittorio Gianni Zaffarana, Matteo Precerutti declare that they have no conflict of interest related to this paper.

Informed consent All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2000 (5). All patients provided (written) informed consent to enrolment in the study and to the inclusion in this article of information that could potentially lead to their identification.

Human and animal studies The study was conducted in accordance with all institutional and national guidelines for the care and use of laboratory animals.

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