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Case Report

Triple locations of elastofibroma dorsi: First case report and review

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ARTICLE INFO

Article history:

Received 8 January 2014

Accepted 29 June 2014

Available online 24 July 2014

Keywords:

Connective tissue neoplasms

Elastofibroma dorsi

Diagnosis

Treatment

ABSTRACT

Introduction: Elastofibroma is a rare benign fibroelastic tumor.

Case: We report a case of a 44-year-old woman, operated 4 months ago for a left infrascapular desmoid tumor and presented for bilateral mass of the left suprascapular and right infrascapular regions which were surgically resected. The diagnosis of elastofibroma was made and also confirmed for the left infrascapular desmoid tumor. Six months later, she presented an asymptomatic left infrascapular recurrence. Monitoring was continued and the patient is still asymptomatic after 24 months.

Discussion: We highlight the importance of imaging features understanding in order to avoid misdiagnosis and unnecessary surgical resection.

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1. Introduction

Elastofibroma dorsi (ED) is a rare benign fibroelastic tumor typically occurring in the periscapular region of elderly patients. These tumors are characterized by slow-growing and frequency of asymptomatic cases. Bilateral ED were frequently reported but cases with triple locations have never been described. Through this report, we discuss the diagnostic and therapeutic management of elastofibroma.

2. Case report

A 44-year-old woman, with no significant medical history, was operated 4 months ago for a left infrascapular chest wall tumor. She had resection and the diagnosis of desmoid tumor with free surgical margins was made. There was no history of manual job or familial similar cases. She presented for a protrusion of the right scapular area. There was no other patient's complaint especially no pain or functional impairment

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<http://dx.doi.org/10.1016/j.jor.2014.06.013>

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Fig. 1 – Axial CT scan images after use of contrast agent demonstrating solid masses, with ill-defined margins, muscular-like density and heterogeneous aspect. a) Right subscapular tumor. b) Left suprascapular tumor.

of the scapular girdle. On clinical exam, there was a bilateral hard mass located in the left suprascapular and right infra-scapular regions. Both tumors were fixed to the deep wall and measured respectively 6×5 and 7×5 cm. The remainder of examination was normal especially no sign of recurrence under the left infrascapular scar or regional lymphadenopathy. Ultrasonography showed oval tumors, with ill-defined margins and inhomogeneous echostructure consisting in alternation of hyper- and hypo-echogenic bands. Doppler not revealed intrinsic vascularization. CT scan demonstrated 2 solid masses, with ill-defined margins, muscular-like density and heterogeneous aspect. The first mass located profoundly within the left trapezius and levator scapulae muscles in contact with the left supraspinous fossa, first three ribs and clavicle. The second tumor located between the subscapular muscle and the 5–8th ribs deep to the serratus anterior muscle. Tumors showed low and heterogeneous enhancement. There was no regional or metastatic extension (Fig. 1). A provisional diagnosis of desmoid tumor was made and surgery was performed. Surgical exploration confirmed radiological features but both tumors were indistinct to bone structures and resection razes ribs. A suction drains was kept inside surgical sites for 48 h. The remainder of the patient's course was uneventful with no loss of motion. Gross pathological examination revealed white tumors, with a rubbery and slightly hard consistency. Microscopically, tumors were poorly circumscribed and showed mesenchymal proliferation associating scattered fibroelastic cells with no malignancy features intermingled with degenerated elastin fibers within an abundant collagenous and myxoid matrix (Fig. 2). The diagnosis of elastofibroma with positive margins in depth alongside ribs plan was made. After re-examination, the diagnosis of desmoid tumor firstly made for the left infra-scapular tumor was rectified to elastofibroma. No supplementary treatment was proposed and the patient was followed up clinically and radiologically. Six months later, she presented an asymptomatic left infrascapular recurrence detected on MRI. The tumor of 5×3 cm was ill-defined with muscular-like signal intensity both on T1- and T2-weighted sequences. The lesion was inhomogeneous and striated and showed moderate enhancement (Fig. 3). Surgery was deferred and we decided to continue monitoring. First clinic and MRI

control was realized at 6 months. Then, the patient was monitored clinically every 6 months with annual MRI exam. After 24-month of periodic assessment, the patient is still asymptomatic with stable tumor size.

3. Discussion

ED occurs usually in elderly individuals with female predilection.¹ Its prevalence is probably underestimated due to frequency of asymptomatic and radiologically undetectable cases.² ED locates mainly in the subscapular region within the serratus anterior muscle.^{3,4} Suprascapular locations are less frequent and may be isolated in half time cases and synchronously associated with subscapular locations in the other half.⁵ Interscapular location has been reported but right side tumors are the commonest.^{5,6} Bilateral ED constitute 25–60% of cases and are often developed asynchronously. The second tumor is frequently discovered fortuitously on clinical exam

Fig. 2 – Histologic findings: Photomicrographs showing scattered fibroelastic cells intermingled with degenerated elastin fibers within an abundant collagenous and myxoid matrix. HES original magnification $\times 400$.

Fig. 3 – MRI study showing the left infrascapular recurrence. a) Axial T1-weighted sequence. b) Axial T2-weighted sequence. c) Coronal T1-weighted sequence after gadolinium injection.

or radiologic imaging.^{3,4,7,8} ED is characterized by slow-growing and spontaneous stop of growth leading to long clinical course and frequent asymptomatic cases.^{2,3,5} Except of their typical location and an evocative bilaterality, ED has unspecific clinical presentation and may be misdiagnosed with others soft-tissue tumors.² To our knowledge, the present case is the first one with triple and bilateral locations of ED ever reported. Its clinical presentation was typical but confusing due to the history of recently resected left infrascapular tumor histologically misdiagnosed as desmoid tumor. MRI is the imaging modality of choice in diagnosing and monitoring of ED.⁹ In the present case, it was efficient to detect post-operative recurrence and exclude fibrotic sequels. Typical clinical and radiological features are often sufficient for a presumptive diagnosis of elastofibroma.² Biopsy is not obligatory but useful in cases of non-operative management especially to exclude malignant tumors.⁷ Asymptomatic ED can be managed conservatively with medical treatment such as non-steroidal anti-inflammatory drugs, associated with manual activity decrease and under clinical and radiological control.^{2,3,7} Actually, there is no consensus concerning modalities of monitoring. The debate concerns particularly the rational of an onerous follow of such slow-growing and little-symptomatic benign tumors.⁷ Overall, an initially close monitoring based on clinical exam and MRI at 3 months is recommended to exclude malignancy and define the tumor growth profile. Then, an annual or biannual control seems to be reasonable.² When ED is painful or responsible for a disabling functional impairment of the scapular girdle, tumors should be resected.⁵ Size tumor and rapidly enlarging are no longer criteria for surgical management. In these cases, biopsy is performed to confirm diagnosis and patients are offered the choice of operative or non operative treatment.^{7,10} Retrospectively, we consider that with the history of resected infrascapular desmoid tumor, it was necessary for our patient to obtain an histological evidence but biopsy would have been probably sufficient to confirm diagnosis and avoid surgical resection. When it is made, surgery must be as complete as possible in order to avoid recurrence. Marginal excisions permit functional sparing but sufficiency of such procedure is controversial because of risk of incomplete removing. Wide or radical excisions are frequently unrealizable because of anatomic constraints.^{7,10,11} In fact, tumors are generally fixed to the ribs plan and as was the case for our patient, resection margins in depth are positive.^{2,5,8} Residual symptoms like

pain and loss in range of motion are uncommon but possible.² There is no consensus concerning post-operative follow-up. We think that the monitoring strategy proposed in cases of conservative management is also relevant after surgery. Local recurrences are rare and always related to incomplete resections.^{5,12} In the present case, resection was probably incomplete but histologically undetected. Recurrences can be conservatively or surgically managed depending on clinical discomfort severity. Use of radiotherapy in case of unresectable and disabling ED is controversed owing to their benignity.¹¹ Prognosis of both resected and conservatively managed tumors, with single or multiple locations, is functionally favorable in addition to inexistent risk of malignant transformation.^{2,5}

4. Conclusion

Elastofibromas are uncommon tumors generally evocative in elderly women. Clinical appearance at diagnosis can be alarming leading to suspicion of soft-tissue sarcoma but subsequent evolution show ungrowing tumors. Multiple locations tumors may occur with no substantial changes in management. We highlight the importance of imaging features understanding particularly on MRI in order to avoid misdiagnosis and unnecessary surgical resection. Conservative management is reasonable to consider in asymptomatic cases and even in symptomatic ones depending on patient preference. When it is made, extent of surgery is the major predictor factor for recurrence. Functional prognosis is favorable even in resected and conservatively managed elastofibroma.

Conflicts of interest

All authors have none to declare.

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