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LETTER / *Senology*

A desmoid tumour associated with a breast prosthesis

F. Gergelé [a,*], F. Guy [b], F. Collin [c], D. Krausé [d]

[a] *Department of Radiology, Bocage Hospital, CHU de Dijon, 1, boulevard Maréchal-De-Lattre, 21000 Dijon, France*
[b] *Department of Radiology, Georges-François-Leclerc Center, 1, rue de Professeur-Marion, 21079 Dijon cedex, France*
[c] *Department of Pathology — Tumor Biology, Georges-François-Leclerc Center, 1, rue de Professeur-Marion, 21079 Dijon cedex, France*
[d] *Department of Radiology, Bocage Hospital, 2, boulevard Maréchal-de-Lattre-de-Tassigny, BP 77908, 21079 Dijon cedex, France*

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A desmoid tumour is a rare tumour occurring predominantly in young adults. It involves deep fibromatosis not giving rise to secondary localisations but having considerable local aggressive potential. The risk factors are still poorly understood. The role of MRI in the initial examination and monitoring is crucial, all the more so since a new therapeutic strategy, with initial monitoring, is undergoing evaluation.

Observation of a case

The main aspect of Mrs Q.'s medical history was an infiltrating ductal adenocarcinoma of the left breast, with invasion of the axillary lymph nodes, discovered when she was aged 38 and in the third month of pregnancy. It was treated by neo-adjuvant chemotherapy during her pregnancy, then by mammectomy and axial lymph node dissection, followed by adjuvant radiochemotherapy. Two years later, she underwent left breast reconstruction with insertion of a prosthesis and restoration of symmetry with a prosthesis on the right.

Five years after diagnosis of the cancer, and 3 years after the reconstruction surgery, a mass appeared within the left breast reconstruction. The ultrasound examination showed a well delineated intramuscular mass, discretely hypoechoic compared with the homogeneous subcutaneous fat. Microbiopsies taken under ultrasound were composed of fusiform cells with elongated nuclei and found to be myofibroblasts with no nuclear abnormality, arranged in bundles on a fibrous base. Immunohistochemical analysis showed recognition of foci of myofibroblasts by anti-actin and anti-beta-catenin antibodies. This indicated a probable desmoid tumour (Fig. 1), which was confirmed by the presence of a gene mutation encoding for beta-catenin.

* Corresponding author.
E-mail address: fgergele@yahoo.fr (F. Gergelé).

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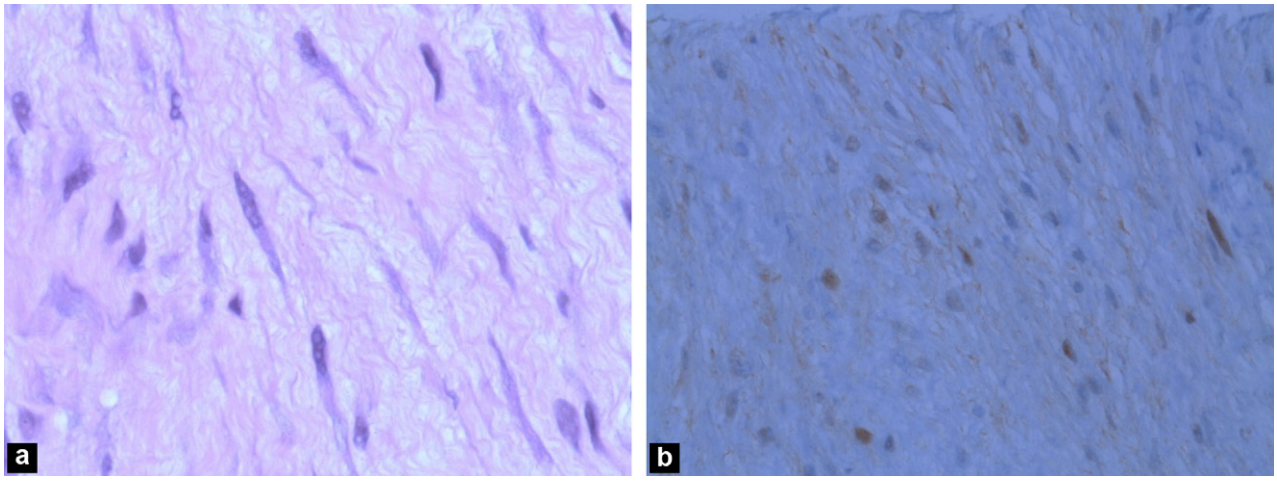

Figure 1. Histological sections of a desmoid tumour: a: fusiform cells with undulating or elongated nuclei corresponding to myofibroblasts, with no nuclear abnormality, arranged in bundles on a fibrous base; b: immunostaining of the myofibroblasts with anti-beta-catenin antibodies.

An initial MRI examination found a well circumscribed mass in the left pectoral muscle, measuring 67 mm high by 48 mm wide, with a large axial diameter of isosignal intensity in T1 weighting compared with the pectoral muscle, a heterogeneous hypersignal in T2 and becoming very weakly enhanced following injection of gadolinium. The prosthesis in contact with the lesion was intact (Fig. 2a,b,c).

Initial MRI and clinical monitoring was suggested. The next check-up at 3 months showed a 25% increase in the major axis of the tumour, i.e. progression according to the RECIST criteria (Fig. 2 d,e,f). Multi-disciplinary consultation resulted in deciding to start treatment with anti-inflammatories and to continue MRI monitoring.

Comments

Desmoid tumours are rare, accounting for 0.03% of all cancers. Their incidence is assessed at two to four per million inhabitants per year [1]. They occur mostly in young adults. They are locally aggressive and tend to recur. Desmoid tumours are deep fibromatoses affecting musculoaponeurotic structures, unlike superficial fibromatoses which affect the fasciae. They are classed in three groups depending on their location: abdominal tumours, which infiltrate the abdominal wall (most often in women during the period of reproductive activity who are using hormone contraceptives, or during or after pregnancy), intra-abdominal tumours, which invade the pelvis, the mesentery and the retroperitoneum (common in Gardner's syndrome) and extra-abdominal tumours which occur in the limbs (70%), the thoracic wall (15%) and the mediastinum. Less than 0.2% of breast tumours are desmoid tumours [1].

The aetiology and how the growth of such tumours is regulated is as yet poorly known. Most of them occur sporadically (97%); there is mutation of the beta-catenin gene in 85% of sporadic tumours. Other tumours occur in the context of Gardner's syndrome, or familial adenomatous polyps (rare 1:1,000,000, an autosomal dominant disease related to mutations of the *APC* gene, chromosome 5q 21-22 with high penetrance, involving polyps of the digestive tract with a potential for malignant transformation, osteomas, cutaneous events and desmoid tumours) [2]. Moreover, the high woman/man ratio (3/1) and the increase in volume of the tumours during pregnancy suggests that hormones are involved, mainly the oestrogens. Surgery and trauma are also risk factors for these tumours occurring. Twenty-five cases of desmoid tumours associated with a breast prosthesis have been reported in the literature; but no link has been demonstrated between the presence of a prosthesis and the occurrence of a desmoid tumour [3]. They occur more often when the prostheses contain silicone rather than saline solution. Approximately 2 years elapse between the implantation of the prosthesis and the appearance of the tumour [3].

The first-line treatment previously was surgery, possibly combined with radiotherapy. However, the surgical results were unpredictable, with a considerable rate of recurrence at 5 years (20 to 60%), and sometimes ruinous as regards function or aesthetics. Several medical treatments have been tested (non-steroidal anti-inflammatory drugs, hormone treatment, interferons, tyrosinase kinase inhibitors and cytotoxic agents) with a variable success rate [1]. Bonvalot's team has shown stability of the disease at 5 years in half the 142 non-treated patients or those treated with drugs [4]. A new strategy is being evaluated consisting of initial monitoring then initiation of drug treatment if the tumour evolves. In the event of failure, surgery, possibly associated with radiotherapy, could be suggested. Studies are underway to understand the biomolecular factors implicated in the evolution of fibromatoses in order to assist in screening patients with an increased risk of evolution, who would then be candidates for surgery.

MRI is the major imaging examination in the initial examination and monitoring. The MRI characteristics of desmoid tumours vary [5]. The tumour is often poorly delineated with isosignal in T1 weighted images relative to the adjacent muscles, is of isosignal or hypersignal intensity in T2-weighted images or STIR, and shows moderate to intense heterogeneous enhancement. Sometimes, the tumour is well delineated in hyposignal in t1 and T2 images with very low enhancement, which indicates a predominantly fibrous

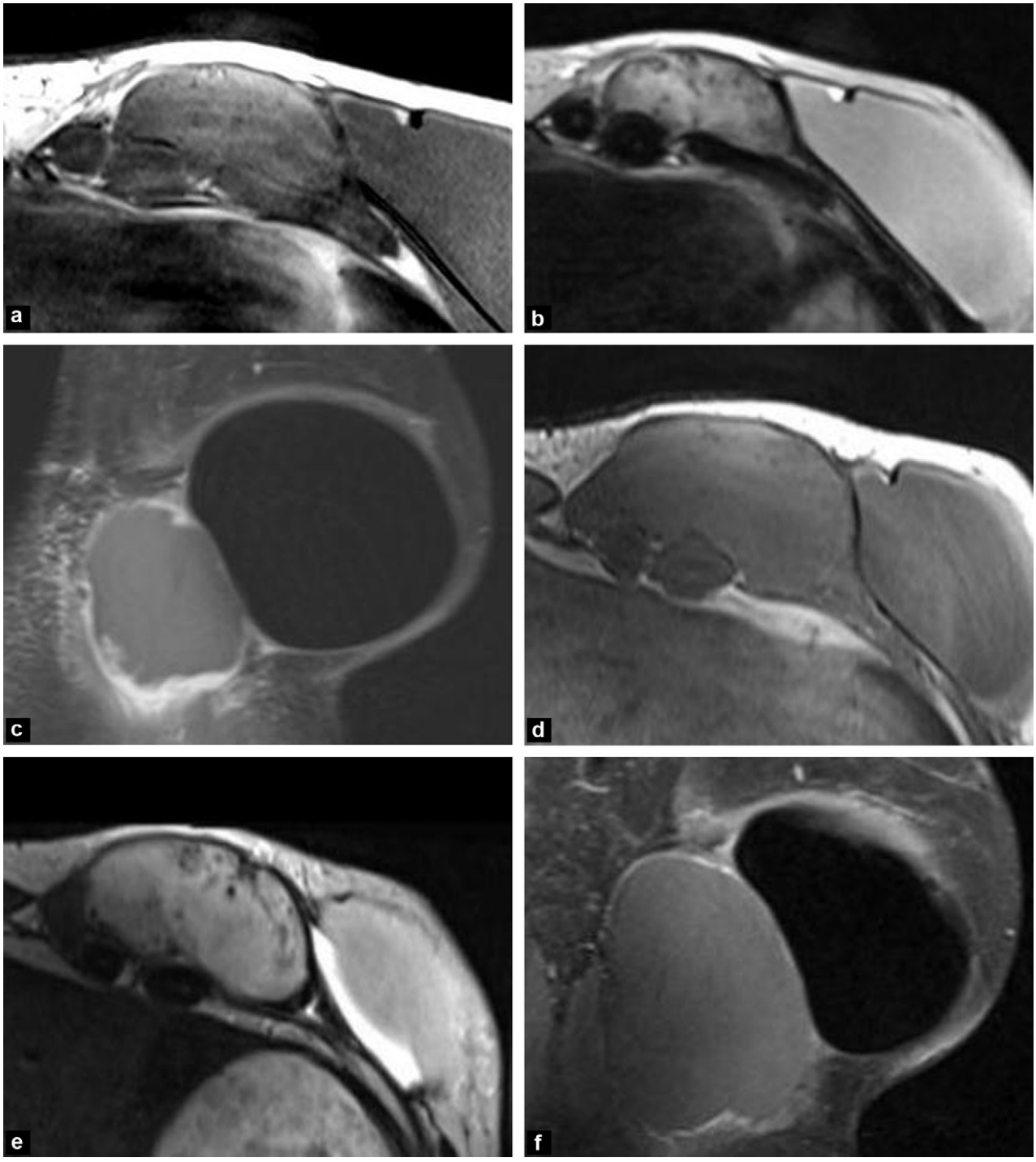

Figure 2. MRI of the left anterior thoracic wall: initial examination (a, b, c) and monitoring examination (d, e, f). Increase in volume of the tumour without modification of the signal: isosignal in T1 weighting relative to the pectoral muscle, heterogeneous hypersignal in T2 weighting and weak enhancement following injection of gadolinium: a: T1-weighted axial slice; b: T2-weighted axial slice; c: T1-weighted coronal slice following injection of gadolinium and fat saturation; d: T1-weighted axial slice; e: T2-weighted axial slice; f: T1-weighted coronal slice following injection of gadolinium and fat saturation.

component. Variation in the T2-weighted signal seems to depend on the cellularity [6]. Certain inconsistent aspects can give a clue to a diagnosis of fibromatosis: hypointense curved areas in T2 weighting (fibrosis septa), the appearance of a hypointense pseudocapsule in T2-weighted images and a hypersignal in T2 weighting less than or equal to that of the subcutaneous fat [7]. The kinetics of enhancement of desmoid tumours studied during dynamic sequences are variable, and seem to depend on cellularity; enhancement of a tumour rich in fibrous tissues is gradual [6,8,9]. No correlation has been demonstrated between the size and the signal: a tumour can increase in size without its signal being

modified. MRI does not allow the evolution of the tumour to be predicted [8]. During monitoring, the efficacy of drug treatment is observed as a reduction in size and an increase in the T2-weighted hyposignal (appearance of fibrous tissue) [10]. Finally, with injection of gadolinium, tumoral recurrence can be distinguished from post-operative fibrosis.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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