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Case Report Open Access

Desmoid Tumors: Three New Observations

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Abstract

Desmoid tumors, also called aggressive fibromatosis, have been described for the first time by John McFarlane in 1832; they are rare, but not exceptional. They are part of deep fibromatosis and described as infiltrating fibrous proliferations without metastasis, but with tendency to recur locally. Their benign histological structure contrasts with their local aggressivity and their therapy remains the major problem. We report three new cases diagnosed in an Internal Medicine Department in three women aged respectively of 54, 27 and 37 years. The respective locations were ovary, inguinal and parietal region of the scapular. Therapy was based mainly on surgery. The desmoid tumor is suspected on clinical and radiological signs and confirmation of diagnosis is pathological. Regular monitoring of patients is necessary because of the frequency of recurrences.

Keywords: Desmoid tumor; Surgery; Radiotherapy

Introduction

Desmoid tumors (desmos = band), also called aggressive fibromatosis, have been described for the first time by John McFarlane in 1832 [1]. They are rare, but not exceptional. They are part of deep fibromatosis and described as infiltrating fibrous proliferations without metastasis, but with tendency to recur locally. These tumors can be located in the abdominal wall, abdomen, or extra-abdominal. Their benign histological structure contrasts with their local aggressivity. Therapy remains the major problem. A better understanding of these tumors would improve their management and their subsequent prognosis. We report three new cases diagnosed in an Internal Medicine Department.

Case Report 1

A woman aged of 54 years, without medical history, was admitted for exploration of a feeling of heaviness in the right flank, epigastric pain and melena evolving one week prior to admission. Clinical examination and biology were normal. Ultrasonography and abdominal CT scan showed a right ovarian mass of 5 cm, with vascular subscapular calcifications. The gastroscopy objectified a gastric ulcer of 7 mm and there were no polyps at colonoscopy. The biopsy of the ulcer did not show malignancy, it was treated medically with good evolution. The family survey in search of similar cases was negative. The patient underwent resection of the tumor whose histology with immunostaining by beta-catenin confirmed its desmoid fibro-sclerotic nature remodeling and calcified. The outcome was favorable and there was no recurrence at twelve months of decline.

Case Report 2

A woman aged of 27 years, without medical history, was admitted for exploration of a left inguinal mass moving six years ago, painful at walking. Clinical examination was unremarkable. Biology was without anomaly. Ultrasonography of the soft tissues showed soft tissue mass of left inguinal of six centimeters long axis. Colonoscopy and gastroscopy didn't objectify abnormalities. MRI confirmed the initial tissue mass left inguinal poorly limited to six centimeters long axis, fairly well vascularized. The patient underwent surgical biopsy of this mass. Histological examination showed a highly vascular tumor, with a fibromatosis consists of a proliferation of mesenchymal cell density made moderate fibroblast-like cell nuclei in fine chromatin and small nucleoli without atypia. Mitoses were very rare. The tumor infiltrated the fatty tissue and glandular structures engainait few. Immunolabeling by beta-catenin was positive. The patient was treated with colchicine and received surgical excision. The evolution was marked by local recurrence one year after the MRI with soft tissue recurrence of locoregional tumor process which was larger and drove back the femoral vascular bundle. Reoperation with postoperative radiotherapy was indicated. The patient was subsequently lost sight of.

Case Report 3

A woman aged of 37 years, having as antecedent treated pulmonary tuberculosis at the age of 17 years, was admitted for exploration of two parietal masses in bilateral scapular moving three years ago, gradually increasing in size and involving joint pain of shoulders mixed type. The examination objectified two parietal masses, sub scapular, bilateral, tissue consistency, movable relative to the superficial surface and adherent to the deep surface. The biological and inflammatory muscle was unremarkable. Ultrasound of soft tissue masses were both objectified oval parietal tissue under scapulars which measured respectively 4.8 and 4.2 cm of diameter. The bulkier sat on the left. These two teams drove back the skin surface and sat directly in contact with the ribs. MRI of the soft tissues of the scapular regions showed that the left mass corresponded to a swollen left big dorsal muscle in subscapular about five centimeters with infiltration of muscle fibers by a fat component without pleuropulmonary lesions suspicious lesion or lung. The right mass was suspicious and resected. Its histology with immunostaining by beta-catenin confirmed desmoid nature. The evolution is stable with a decline of 48 months.

Discussion

Desmoid tumors (DT) are rare, they correspond to 0.03% of all neoplasms, with an incidence of 2 to 4 new cases per 100.000 habitants [1,2]. They occur sporadically or as part of Familial adenomatous polyposis (FAP) [3]. The frequency of DT associated with FAP varies

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according to authors of 4% to 32% [3]. Contributing factors for DT are trauma or abdominal surgery in 75% of cases, particularly after prophylactic colectomy [2]. The DT appears on average in the three years following the initial surgery, often within five years after colectomy [4]. Hormonal factors are implicated; in fact, the incidence of DT is higher in women of childbearing age, the case of our second and third observations, during pregnancy or after childbirth, but also at the taking of oral contraception. *In vitro*, cells from desmoid tumors in the presence of estrogen have multiplied and produced collagen while their growth was inhibited by anti-estrogens [1-3,5]. Genetic factors are also involved in the pathogenesis of DT, in fact, the FAP is an autosomal dominant inherited disorder linked to mutations in the adenomatous polyposis coli tumor suppressor gene, which is a tumor suppressor gene located on the chromosome 5. Thus, from 10 to 20% of patients with the mutation develop a DT. This affects women twice, with an average age of onset of 30 years [3]. Thus, it is essential to perform genetic research with families of patients with DT [6]. The association DT, FAP, osteomas, sebaceous cysts of the face is Gardner's syndrome [3,5]. The clinical relevance of DT is poor. The extra-abdominal fibromatosis occur preferentially between puberty and the age of 40 years with a peak frequency between 25 and 35. Women are two times more affected than men [3]. The tumor appears as an ill-defined mass, slow growth, little or no sensitive. Discomfort to mobility may occur depending on the location and extent of the tumor. The extra-abdominal locations are rarely multifocal [3]. Wall abdominal DT are palpable masses, most often at the rectus and external oblique of the abdomen. Their size varies from three to ten centimeters in diameter and they never cross the midline. They occur mainly in young pregnant women, or more frequently in the first year after pregnancy [3,6]. The mesenteric DT is the most common tumor of the mesentery, it is most often sporadic, but 10% of cases are parts of Gardner's syndrome. It comes at a very variable age, it affects both men and women, it is most often localized in the mesentery of the small intestine, but with ileo colonic mesentery, the omentum or the retroperitoneum were described. The discovery can be done at the balance sheet of an increase in abdominal girth. However, this tumor may be incidentally during a laparotomy or when a complication (bowel obstruction, gastrointestinal fistula, signs of ureteral compression, mesenteric ischemia, deep vein thrombosis, compression causing neurological deficits or dysfunction of the ileal reservoir) [3,4]. On X rays, the lesion appears as a soft tissue mass affecting breaking plans Inter underlying muscle. There is often a contiguous bone reaction [1]. The abdominal CT is the modality of choice for diagnosis and performs surveillance [2,3,7]. MRI has better resolution than the scanner to differentiate between postoperative alterations of a DT after medical treatment of a recurrence. Moreover, it would be possible by this examination to predict tumor growth since T2; a hyper-signal is associated with a high cellularity [1,3]. The CT and MR angiography can clearly identify the relationship of the tumor with blood vessels and make the differential diagnosis with fibrosarcoma (poverty of desmoid tumor vasculature compared to the significant neovascularization of poorly differentiated sarcomas) [6]. Colonoscopy can search polyposis colorectal upon detection of a desmoid tumor [8,9]. The gastroscopy is useful to look for polyps or gastric ulcers. Indeed, one can attend a desmoid tumor associated with FAP with no polyps in the colon. It is then necessary to know the look in the upper digestive tract [8]. Pathological examination can range from a simple fibrous lesion in fibrosarcoma [10]; the macroscopic appearance of the DT is usually confined to the level of the fascia and the muscle fascia, which can infiltrate the subcutaneous tissue. The tumor is firm, cut in granite, of a shining white, coarsely trabeculated resembling scar tissue. The microscopic appearance of the tumor is generally ill-defined with infiltration of adjacent tissues. Proliferation is in the form of elongated cells, fusiform, small size, uniform in appearance, separated from one another by abundant collagen tissue non inflammatory. Cells have a small kernel, without atypia. They set vimentin and muscle actin, calcifications are rare. Immunolabeling by beta-catenin and Ki67 is positive and confirms the diagnosis [3]. Desmoid tumors are usually slow growing. In 10% of cases they regress spontaneously (especially after menopause) [5]. The local recurrence rate exceeds 90% [3]. The risk of recurrence would be even higher than it would for women aged below 30 years, forms of extra-abdominal and distal, was seen in our second case. The strategy supported depends on the resectability of the tumor. It can be a simple oversight. Indeed, symptomatic uncomplicated DT can be surgically resected, if it is small, well defined and without invasion of vital structures, the case of our observations. Macroscopic resection is possible in two thirds of cases, but it is difficult for the surgeon to be sure of having a complete resection because the tumor is not encapsulated [3]. Surgical treatment with curative intent should remain the treatment of choice, although it does not prevent the risk of recurrence. Radiotherapy provides a definite advantage when residual disease or inoperable sites [3]. Series have shown a profit, with 6% relapse versus 28% without radiotherapy [9]. For the wall desmoid tumors, radiation therapy has proven effective in reducing the recurrence rate [1-2]. Recent studies have shown that when the tumor site allows, radiotherapy alone gives similar results to surgical resection [1]. In case of relapse or invaded margins, adjuvant external beam radiotherapy improves local control (25% with, versus 59% without) [3]. If macroscopic tumor is inoperable, it also allows in 78% of tumor control [3]. In addition, DT grows slowly, which implies a low chemosensitivity. The frequency of occurrence of DT in women, the presence of estrogen receptors and regression at menopause give hope for hormone sensitivity. So, anti-estrogens were tested such as tamoxifen, GnRH agonists and progestins. Finally, it is through their anti cox2, by inhibiting prostaglandin synthesis, which stimulates the immune response against tumor that NSAIDs act. Both products were used, sulindac and indomethacin. The recommended dosage is 120 mg/day for indomethacin and 300 mg/day sulindac [3]. The current standard of treatment is to attempt NSAIDs first, then SERMs (Selective estrogen receptor modulators), and only to consider surgery for symptomatic complicated patients [11]. Other studies reported the positive role of isolated limb perfusion especially for irresectable extremity desmoids tumor [11-13]. Certainly, treatment of patients with desmoids tumors should include a multidisciplinary approach, in a clinical trial setting, with collection of fresh tissue and serum for future studies [14]. Our patients did not receive preoperative radiotherapy or tamoxifen or suldinac because these therapies were not immediately available and patients well advanced in post surgery.

Conclusion

The desmoid tumor is a rare benign tumor. It is suspected on radiological data, but confirmation of diagnosis is pathological. Regular monitoring of patients is necessary because of the frequency of recurrences.

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