Aggressive angiomyxoma of the pelvis: a clinicopathologic study of a case

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Summary

A case of aggressive angiomyxoma in a 38-year-old pregnant woman is described. These clinicopathologically distinctive soft tissue tumors comprise a quite rare entity. They are usually located in the pelvic or perineal soft parts and occur predominantly in women. On gross inspection, these lesions are typically soft bulky masses, frequently with a multilobulated contour and of gelatinous appearance. They are characterized by slow growth, infiltration of adjacent structures, absence of distant metastases and a tendency to recur locally, probably secondary to incomplete excision. Therefore, treatment should consist of wide surgical excision, as complete as technically possible.

This is the second case of aggressive angiomyxoma of the pelvis occurring in a pregnant woman.

Key words: Mesenchymal tumors of the pelvis; Myxoma; Angiomyxoma.

Introduction

Myxomas were initially described by Virchow in 1863 [1]. Ever since, there has been remarkable progress in the classification of this type of soft tissue tumor so that today we can speak more accurately about the malignant behaviour or the prognosis of the various cases [2, 3]. Steeper and Rosai were the first to report a distinctive type of myxoma with unique histologic characteristics and biological behavior [4]. These tumors have since been described as angiomyxomas. They predominantly involve the pelvis and perineum of young women and are characterized by their diffuse myxoid appearance, the presence of neoplastic blood vessels, their locally infiltrative nature and their tendency for multiple local reccurences. The aim of the present study was the presentation of a pelvic angiomyxoma case as well as a short review of the international literature.

Description of the case

1. Clinical and laboratory data

A 38-year-old woman who underwent a caesarian section about a year ago, presented with a soft palpable extraperitoneal swelling in the right lateral part of the pelvis. The initial clinical impression was that of a regional haematoma. Follow-up of the mass with ultrasound and computed tomographic scan revealed a progressive increase in size over a two-month period and eventually exploratory laparotomy was performed.

2. Operative findings

The patient underwent exploratory laparotomy. At surgery, a retroperitoneal, soft in consistency (15x12x6 cm) mass was found filing the right lateral part of the pelvic cavity displacing medially the right ureter, the uterus and bladder. Its lower part was in close relation to the pelvic floor. The tumor proved to be markedly soft and friable, easily separated from the surrounding structures. Excision was carried out without disruption of the lesion capsule by sharp dissection with diathermy. The right ureter and the great vessels of the pelvis were identified and carefully protected. The postopoerative course was uneventful and there has been no evidence of reccurrence to date (5-year follow-up).

3. Pathological findings

The excized tumor measured 15x12x6 cms. The external surface was lobulated with finger-like projections and grossly gave the impression that it was surrounded by a capsule. The cut surface was homogenous, gelatinous and focally microcystic. The tumor consistency was firm.

Histologically the tumor presents a myxoid eosinophilic background containing numerous vessels of varying size, focally clustered (Fig. 1). The neoplastic cells are spindle shaped with round or oval dark nuclei (Fig. 2). No significant cellular atypia or mitotic activity can be observed. The circumference of the tumor is focally rich in collagen fibres, giving the impression of a pseudocapsule which is infiltrated by the neoplastic cells.

To define the nature of this tumor, an immunohistochemical study was undertaken on formalin-fixed, paraffin-embedded tissue sections, for the following antibodies:

Vimentin (Novocosta Monoclonal), Factor VIII (Dako, Polyclonal), Desmin (Sigma, Polyclonic), S100 (Immunon, Polyclonal), A1-Antitrypsin (Dako, polyclonal): All the antibodies were stained with astreptavidin-alcaline phosphatase amplification system (Biogenex Super sensitive kit). Hormonal receptor status was determined by a Novocastra NCL-ER/PRG Paraffin kit, which employs an ABC technique, carried out according to manufacturer's instructions.

Tumor cells gave an extensive positive reaction for vimentin and a focally positive reaction for desmin. There was a

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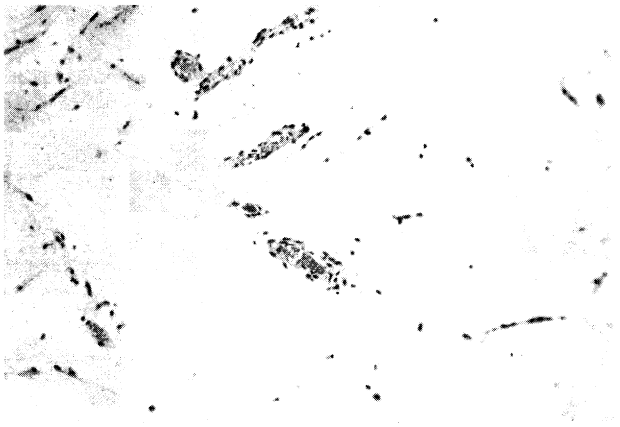

Figure 1. — Aggressive angiomyxoma of the pelvis. Histological section showing hypocellular area with myxomatous background and prominent vessels (H-Ex150).

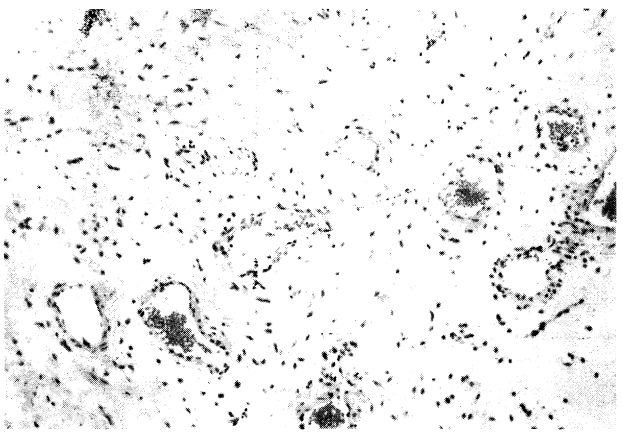

Figure 2. — Aggressive angiomyxoma of the pelvis. Histological section showing spindle cells and "thick-walled" vasculature empedded in a loose myxoid stroma (Haematoxylin-eosin stain x150).

negative reaction to S100, A-1-antitrypsin and Factor VIII which stained the endothelial lining of the vessel walls. Neoplastic cells were negative for either estrogen or progesterone receptors.

In conclusion, the location of the tumor, the pathological features and the immunohistochemical characteristics, the absence of mitotic activity, lipomatous or neural differentiation, are consistent with the diagnosis of aggressive angiomyxoma.

Discussion

In 1863 Virchow was probably the first to recognise myxomas as a distinct clinicopathologic entity. The same investigator observed that tumors describe as "myxomas" and located in the female pelvic soft tissues, displayed a locally infiltrative behavior with a tendency for multiple recurrences [1]. Occasional case reports and series of large tumors of the female pelvic floor or myxoid neoplasms which had attained giant proportions can be found in the surgical literature [5, 6, 7, 8].

Steeper and Rosai identified the histologic characteristics of this distinct group of soft tumors and invented the term aggressive angiomyxoma to emphasise the neoplastic nature of the blood vessels and its locally infiltrative and recurrent nature [4].

Angiomyxomas are rare forms of neoplasms that tend to occur more frequently in young women with a mean age of 34 years while they are uncommon in men [9]. They are located in the female genitalia and pelvic soft tissues and the sites of involvement are typically described as vulvar, perineal, paravaginal or pararectal. Our case is the second one in the literature reporting this tumor a pregnant woman [10].

The negative immunohistochemical reaction for hormonal receptors in our case support the theory that this tumor is hormone-independent, but the number of cases examined is inadeguate to reach valid conclusions.

On gross examination these tumors are characteristically large, soft masses with their external surface partially or completely encapsulated. Their size varies widely and can attain gigantic proportions [4, 5]. The cut surface usually reveals a gelatinous, white-gray tumor of homogeneous consistency. Histological criteria necessary for diagnosis include spindle-shaped or stellate cells widely distributed in a loose myxoid stroma. Varying degrees of cystic regeneration can occasionally be seen while the lesion displays a well-developed vascular component with vessels of varying size. There is virtually no mitotic activity in order to reach a valid conclusion [4].

A wide variety of benign and malignant mesenchymal tumors must be considered in the differential diagnosis, such as the angiomyofibroblastoma, which is a benign tumor, with a well-defined capsule, prominent cellularity and no clustered vessels; the myxoid liposarcoma, showing a characteristic vascular pattern and lipoblasts; and, the myxoid malignant fibrous histiocytoma, which presents a prominent storiform pattern, cellular atypia and mitotic activity as well as a positive expression of a-1-antitrypsin. Some neurogenous tumors may present a myxoid pattern as well, but invariably show a strong positive reaction to S100 antibody [4, 5, 10].

There is no evidence of distal metastatic spread, but angiomyxomas display a locally aggressive behavior and a high potential for local recurrence [4, 11]. Infiltration of adjacent structures may not always be ruled out during surgery. In view of the above, wide excision and strict follow up appears to be the best treatment.

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