Angiomyxoma diagnosed in a man presenting for abdominal lipectomy

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Aggressive angiomyxoma is a rare tumour that is locally infiltrative but nonmetastasizing. It occurs nearly exclusively in adult women of childbearing age and almost always arises in the perineum and pelvic area. A case of angiomyxoma occurring in a middle-aged, morbidly obese man is reported. The clinical presentation was one of progressing scrotal edema and enlargement with subsequent development of scrotal abscesses, requiring several incisions and drainages as well as oral and intravenous antibiotics. His symptoms and body habitus left him in a significantly debilitated state, prompting him to seek treatment for his conditions. He initially presented for abdominal lipectomy. However, due to the chronic scrotal infections and enlargement, scrotoectomy was recommended before any other surgical procedures. A scrotoectomy was performed by the urology service. Pathological diagnosis of the excised tumour was an aggressive angiomyxoma. Given the rarity of this tumour, especially in males, the proper diagnosis and treatment of angiomyxoma is still being investigated. With this in mind, the physical signs and symptoms that characterize this condition are presented and treatment options are reviewed.

Key Words: Aggressive; Angiomyxoma; Male

CASE PRESENTATION

A 47-year-old morbidly obese man presented requesting abdominal lipectomy. His past history was significant for progressive weight gain to 204 kg, with a gain of 124 kg over a 20-year time span (Figure 1). His obesity led to several comorbidities and treatments including chronic infections, tracheostomy for hypoventilation syndrome causing sleep apnea, and an indwelling perineal catheter for urinary drainage.

Upon referral to the plastic surgery service for abdominal lipectomy, it was decided that before any abdominal lipectomy could be considered, the patient required a scrotoectomy due to his chronic infections and massive lymphedema of the pelvic region and lower extremities. He was started on intravenous antibiotics by the infectious disease service approximately two weeks before this surgery. Evaluation by the urology service included ultrasound and x-rays. Physical examination of his testicles found normal architecture of the testicles and no bowel within the scrotum. No identifiable tumour was diagnosed at that time, and the patient underwent scrotoectomy with approximately 17 kg of tissue removed from the scrotum (Figure 2). He tolerated the procedure well and was discharged back to the Veterans Administration Hospital on postoperative day 2.

Gross description of the specimen was reported as a mass of soft tissue and overlying skin weighing 16.8 kg, markedly edematous, with clear serous fluid. No obvious cysts, hemorrhagic or

Angiomyxome diagnostiqué chez un homme vu pour lipectomie abdominale

L'angiomyxome agressif est une tumeur rare caractérisée par une infiltration locale sans métastases. Il s'observe presque exclusivement chez des femmes adultes fertiles et se manifeste presque toujours au niveau du périnée et de la région pelvienne. On décrit ici un cas d'angiomyxome chez un homme d'âge moyen atteint d'obésité morbide. Le tableau clinique en est un d'œdème et d'hypertrophie progressifs du scrotum avec abcès scrotaux subséquents nécessitant plusieurs incisions et drainages, de même que des antibiothérapies orales et intraveineuses. Sa symptomatologie et sa morphologie problématiques ont amené le patient à consulter. Au départ, il souhaitait subir une lipectomie abdominale. Par contre, en raison de ses infections et hypertrophies scrotales chroniques, une scrotoectomie a été recommandée avant toute autre intervention chirurgicale. La scrotoectomie a été réalisée par le service d'urologie. Le diagnostic anatomopathologique de la tumeur excisée a été un angiomyxome agressif. Compte tenu de la rareté de cette tumeur, surtout chez les hommes, le diagnostic et le traitement adéquats sont encore à déterminer. Cela dit, les signes et symptômes physiques caractéristiques de cette maladie sont présentés de même que les options thérapeutiques envisageables.

necrotic areas were identified. Microscopic description found a mesenchymal lesion composed of myxoid tissue with spindle and stellate cells, without significant atypia or neoplastic cells. Scattered throughout the myxomatous background were several blood vessels of varying sizes. Diagnosis of the specimen was that of a myxoid stromal tumour with features consistent with aggressive angiomyxoma.

DISCUSSION

Aggressive angiomyxoma was originally cited in 1983 by Steeper and Rosai (1), who described this uncommon soft tissue tumour of the pelvis or perineum, arising in premenopausal women. This tumour was given its name due to the typical pathological findings of stellate and spindled cells along with variable-sized blood vessels intricately entwined within a myxoid matrix. Despite their cytologically insipid appearance, these tumours have a high risk of local recurrence. However, they have no metastatic potential.

Aggressive angiomyxoma is a rare, locally aggressive tumour characterized by enlarging, nonpainful masses in the pelvic or perineal regions, almost always seen in young women of childbearing age (95% female predominance). There have been very few reports describing this condition, and our literature search found fewer than 20 reported cases in men worldwide (2-8).

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Figure 1) Preoperative appearance of a 47-year-old man with scrotal angiomyxoma

Benign mesenchymal neoplasms have been linked to an abnormal DNA binding protein. Like numerous benign mesenchymal tumours, this locally aggressive tumour is associated with rearrangements near or within high-mobility group (non-histone chromosomal) protein isoform I-C on chromosome 12, but investigation is still underway (9). Studies have shown that aggressive angiomyxoma in women expresses estrogen receptor/progesterone receptor positivity, thus it is possibly a hormonally responsive neoplasm (10).

The pathological features of aggressive angiomyxoma in men are similar to the cytological findings described in women. Clinical features in men are similar to those in women and have a predilection for the pelvis and perineum. The physical presentation in men is also consistent with the typical presentation in women (11). The men in cited cases have all been middle-aged, and presented with asymptomatic masses in the genital area (2-8).

Differential diagnosis in women includes Bartholin gland cysts or perineal hernias, while in men, aggressive angiomyxomas have frequently been mistaken for inguinal hernias (12). Pathologically, the tumour should be distinguished between a large spectrum of tumours with low-risk local recurrences and those with high-risk metastatic potential. The former includes myxoid neurofibroma, intramuscular myxoma, myxoid lipoma and angiomyofibroblastoma, while those of the latter include myxoid liposarcoma, malignant fibrous histiocytoma and rhabdomyosarcoma.

The treatment of aggressive angiomyxoma is surgical. Wide excision of the tumour with negative margins has shown no recurrence. However, incomplete excision of the tumour has shown a 30% recurrence rate, and thus, meticulous dissection of the tumour should be undertaken by the surgical service. Although the relative rapid growth of these tumours initially raise suspicion for a malignant cause, immunohistochemical staining showing the bland cells typical of aggressive angiomyxoma, as well as the histological features described previously, serve to confirm the diagnosis.

SUMMARY

Aggressive angiomyxoma is a rare, locally aggressive, non-metastasizing tumour that occurs nearly exclusively in the perineal and pelvic region of women of childbearing age. The pathological and clinical features in men are similar to those

Figure 2) Postoperative appearance after resection of the scrotal angiomyxoma

found in women. Presentation in men occurs in the third to fifth decade of life, and is characterized by an asymptomatic, rapidly enlarging mass in the genital area. Suspicion should arise in men with such a presentation when physical examination does not diagnose any of the more common anomalies found in the perineal area, and tissue biopsy should be obtained. Although few reported cases of aggressive angiomyxoma in men have been reported in the literature, it should be considered in the differential diagnosis of rapidly enlarging masses in the scrotal or perineal regions of men. Surgical excision is warranted.

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