CASE REPORT

Uncommon perineal tumours: caution with aggressive surgical management

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SUMMARY

An asymptomatic 66-year-old woman showed a large perineal mass extending close to pelvic organs on MRI. CT-guided needle biopsies revealed a desmoid tumour (DT). The patient refused radical surgery. Four years later, the tumour had marginally increased in size and was still asymptomatic. The revision of earlier biopsies then revealed typical aspects of aggressive angiomyxoma (AA). AA and DT are rare mesenchymal tumours of low-grade malignancy, usually of large size, that occurs in female pelvi-perineal region. Radical resection with wide margins is classically advocated in such tumours in order to prevent the high risk of recurrences. However, due to a slow growth, rare infiltration of adjacent organs and a very low metastatic potential, a watchful waiting policy can be proposed when high postoperative morbidity is expected. In order to propose the accurate treatment, frontline biopsies of the tumour are essential.

radical surgery and was proposed a watchful waiting policy.

OUTCOME AND FOLLOW-UP

After 4 years of follow-up, the tumour remained clinically asymptomatic. The patient only described occasional discomfort during sporting activities but did not relate any pain. MRI showed a marginal increase in size over the follow-up period (104×56 mm; figure 1C). Slides from the first histological analysis were reviewed and an additional immunohistochemical analysis was performed (figure 1D). Microscopically, the tumour was myxoid, with a poor component of spindle cells, without cellular atypia or mitosis. Thick and thin-walled variably sized vascular channels were seen. The phenotype of the tumour cells was: muscle-specific actin+, desmin–, PS100–, oestrogen+, progesterone+. Finally, we concluded with the diagnosis of AA.

BACKGROUND

Perineal tumours have various appearances according to their histological origin and malignancy. Low-grade malignancy tumours, such as aggressive angiomyxoma (AA) or desmoid tumours (DTs) appear as large masses that displace rather than invade local organs. Their good prognosis raises the problem of the appropriate therapeutic decision, especially in large tumours that require perineal surgery of high morbidity. Though complete surgical resection remains classically advocated in all uncommon perineal tumours, a conservative approach can be proposed in selected cases. Our case report illustrates this alternative therapeutic option and its indications.

CASE PRESENTATION

A 66-year-old woman presented with a perineal mass since months. Her medical history was marked by a hysterectomy, a transobturators tape procedure and a rectopexy. Her single complaint was aesthetical. The clinical examination showed a firm mass located in the right ischio-rectal fossa (figure 1A). The digital vaginal and rectal examination confirmed the presence of a mobile tumour without any intraluminal component. MRI revealed a homogenous tissular mass of 88×44 mm, hypointense on T1 sequences. On T2 images, the hyperintense signal intensely enhanced after intravenous contrast (figure 1B). The tumour displaced the vagina and was interposed between the rectum and the urethra without invading them. CT-guided needle biopsies were performed. Histological analysis first described a DT. The patient refused

DISCUSSION

The management of perineal tumours such as sarcoma or anal canal squamous cell carcinoma is challenging. As complete radical resection with wide margins is the basis of curative treatment, resultant major morbidity is frequent. Indeed, the risk of stoma and urogenital tract damages threaten postoperative quality of life. In these last 20 years, improvement in histopathological techniques and pathology description permitted to describe new neoplasms of lower malignant grade as AA or DT (also called aggressive fibromatosis). The 'aggressive' designation refers to their local infiltrative potential and recurrent nature. However, these tumours are characterised by a slow growth and almost non-existent metastasis. AA and DT classical treatments consisted of a radical excision with wide margins in order to limit the high risk of recurrence. Although this management achieved good results in few series,^{1 2} it is today debated. Some retrospective studies reported similar recurrence rates in patients with wide margins compared with tumour-involved resection margins.^{3 4} Moreover, as enhanced by our patient, AA and DT natural histories are marked by a succession of slow increase or stable phases. Therefore, aggressive perineal surgery is not systematically justified in such tumours with excellent prognosis.⁵ On the contrary, several authors proposed frontline watchful waiting policy in perineal AA and DT with predictable resultant morbidity until symptoms or increase in size occur.^{1 6} In conclusion, the optimal management of uncommon perineal tumours requires

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Figure 1 Clinical, MRI and histopathological aspects of a large perineal aggressive angiomyxoma. (A) At clinical examination, a perineal renitent swelling was seen. (B) At diagnosis and (C) 4 years later, respectively, contrast-enhanced T2-weighted MRI showed a large hyperintense tumour (arrows), interposed between the rectum and the urethra. The tumour size marginally increased in 4 years. (D) Microscopically, the tumour was myxoid with a poor component of spindle cells.

accurate biopsies prior to any treatment initiation. Histological diagnosis is crucial to detect these low-grade malignant tumours that are not always to be resected in first line, on the contrary to other aggressive lesions.

Learning points

- Large perineal atypical masses are not always of poor prognosis and can reveal tumours of low-grade malignancy such as desmoid tumours or aggressive angiomyxoma.
- Tumours of low-grade malignancy are not systematically to be resected in first line when a high postoperative morbidity is expected. They can be managed by a watchful waiting policy until symptoms and/or fast increase in size appear.
- In atypical perineal tumours, accurate biopsies must be realised prior to any treatment initiation in order to avoid aggressive surgery that is not systematically indicated in first line.

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contributed to manuscript review. NR contributed to patient management and follow-up. GM recruited the patient, collected the clinical data and contributed to manuscript review.

Competing interests None.

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