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Desmoplastic fibroma of the ilium☆

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ABSTRACT

INTRODUCTION: The desmoplastic fibroma is a rare locally invasive bone tumour. Surgical resection with minimal margins is recommended.
PRESENTATION OF CASE: A 15 year-old boy was referred with chronic left thigh pain. MRI revealed a bone lesion within the cavity of the inner table of the left iliac wing without invasion of the underlying bone marrow. A surgical biopsy revealed a desmoplastic bone fibroma. A partial resection of the inner table of the iliac wing sparing the outer table was performed. At the latest follow-up the initially spared iliac wing had needed further resection. The reason proposed for this is devascularisation by substantial periosteal stripping causing partial resorption initially, then necrosis and ultimately ulceration through the skin necessitating further surgical resection.
DISCUSSION: The technique of resection of a pelvic desmoplastic fibroma sparing the outer table of the iliac wing has not previously been reported. The objective of a limited resection was to minimize the risk of a postsurgical limp caused by weakness of the gluteus medius muscle. However we report that this technique did not work in this case. A wider resection of the iliac wing as it is recommended for a malignant tumour would have yielded a similar final outcome.
CONCLUSION: A partial resection of the iliac wing seemed an appealing technique for a benign tumour of the inner table of the iliac wing. However, considering the complications encountered, the authors advise a simple "en bloc" resection of the iliac wing for this type of tumour in this location.

1. Introduction

Desmoplastic fibroma is a rare benign bone tumour, described by Jaffe[1] in 1958.

It represents 0.06% of all osseous tumours and 0.3% of benign osseous tumors[2–7]. It occurs during the first three decades of life in over 75% of cases, with an equal male and female preponderance[2].

Pain, swelling or more rarely pathological fractures are the most frequent presenting symptoms[2,8,9]. The mandible, femur, pelvis, radius and tibia are the most common reported sites[2,5–7], although every bone of the peripheral skeleton can be affected[10].

The desmoplastic fibroma is a slowly progressing locally invasive tumour, characterized by a production of collagen fibres by its tumour cells. A sarcomatoid transformation is unusual[2,11]. The rare cases which have been described are considered as an underestimate of the actual prevalence of this tumour[10]. The average delay for recurrence is about three years according to Bohm[2].

It has been reported that intralesional surgical procedures such as curettage have a high rate of recurrence, which has been estimated as between 50% and 72%[2,12,13]. Extralesional resection procedures seem to give better results with only a 5% recurrence[2]. Thus "en bloc" surgical resection is recommended.

We report the case of an "en bloc" resection of a desmoplastic bone fibroma of the iliac wing by partial resection of the inner table of the iliac wing. The pros and cons of this technique in this rare location of desmoplastic fibroma are discussed.

2. Case report

A 15 year-old boy was referred for investigation of chronic left thigh pain. The initial clinical examination was normal but an X-ray showed an osteolytic lesion of the left iliac wing.

MRI and CT-scan revealed a well-defined bone lesion (5 cm × 8 cm × 2 cm) within the cavity of the inner table of the left iliac wing without invasion of the underlying bone marrow (Figs. 1 and 2). A Tc 99 bone scan revealed no other lesion. A surgical biopsy diagnosed a desmoplastic bone fibroma.

A multidisciplinary meeting recommended a partial resection of the inner table of the iliac wing. After exposing the outer and inner iliac fossae, the iliac crest was resected with a Gouge chisel. A progressive curettage of the cancellous bone created a gap between the

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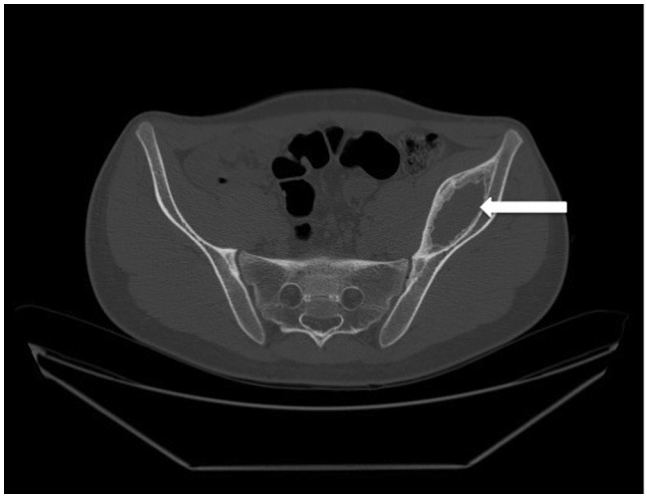

Fig. 1. Frontal T2 weighted MRI showing a tumour formation on the internal part of the left iliac wing.

two tables. Then the tumour was lifted progressively with a Cobb elevator. Periosteum and overlying muscles were then sutured to the outer table. The macroscopic and microscopic findings are illustrated in Figs. 3–5.

A haematoma formed both anteriorly and posteriorly to the iliac wing two days postoperatively but did not necessitate drainage. Three months after surgery, X-rays showed partial resorption of the posterior part of the remaining iliac wing. Six months after surgery a 3 cm skin lesion appeared with bone exposure of the iliac wing and required a secondary resection of the bone sequestrum.

At two years follow-up, the patient's scar has healed and the local skin ulceration has not recurred. MRI did not show local relapse. The X-Ray at last follow-up shows the resorption of the iliac wing (Fig. 6). The patient walks without crutches but with a slight Trendelenburg gait.

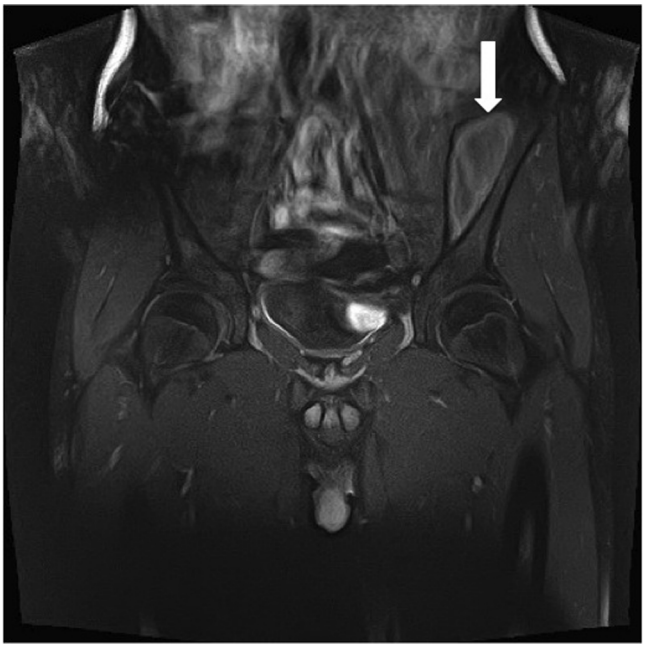

Fig. 2. Coronal CT scan showing a well-limited bone lesion within the cavity of the inner table of the left iliac wing without invasion of the underlying bone marrow.

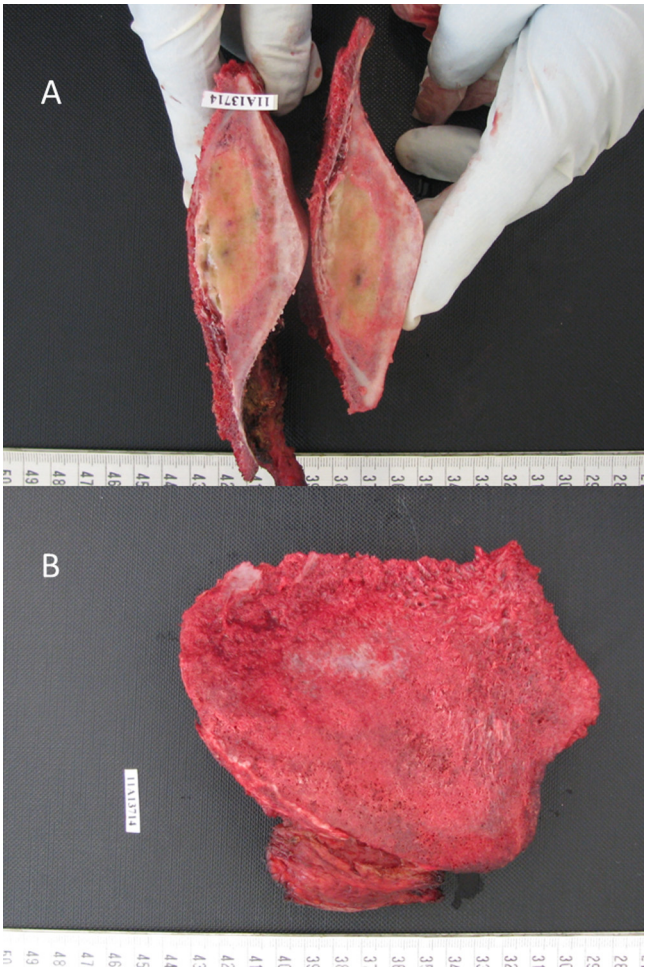

Fig. 3. A: Photograph showing the internal part of the tumour once split in two pieces. The desmoplastic bone fibroma is macroscopically a firm, elastic, homogenous whitish tissue resembling rubber. B: Photograph showing "cancellous bone" side of the tumour.

3. Discussion

Desmoplastic fibroma is a benign bone tumour but is locally invasive.

X-ray usually shows a lytic, honeycomb lesion, with no mineralized matrix and a well circumscribed border[2,5,8,14]. MRI shows the heterogeneous histologic character of the tumour. MRI is the "gold standard" to define the local extent of the tumour before surgery as well as for post surgery follow-up to look for recurrence of the tumour[7,10,15]. Surgically, it has been reported that there is a clear separation between the tumour and the adjacent bone tissue[10]. However, histologically, there are very thin, intracortical tumour strips[16] (Fig. 4). These findings explain the extremely high rate of recurrence when intralesional surgical excision is performed. Because of the connection between tumoral tissue and the adjacent bone, extralesional resection including peritumoral bone tissue is recommended[2,4,6,17].

In the case presented here, a CT-scan and an MRI scan showed that the tumour was exclusively at the expense of the inner table of the iliac wing with no invasion of the soft tissue and the outer table. Taking into account the nature of the tumour, it was decided to conduct an extralesional resection sparing the outer table of the iliac bone with minimal surgical margins. The theoretical objective was to minimize the risk of postsurgical Trendelenburg limping caused by weakening of the gluteus medius muscle as is observed in complete resection of the iliac wing. The rationale in preserving

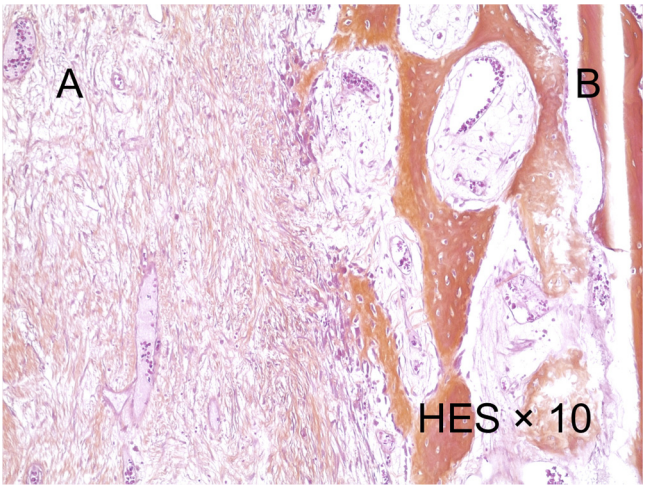

Fig. 4. Photograph showing the microscopic findings at the junction between the tumour (A) and the iliac wing (B). There is no clear junction between normal bone and the tumour. There are very intracortical tumour strips, which explain why a simple curettage is not recommended.

the outer table was to enable the iliacus and gluteus muscles to adhere to the remaining iliac crest.

However, follow-up X-ray showed that the posterior part of the outer table resorbed spontaneously 3 months after surgery. The necrotic bone was spontaneously eliminated through the skin as a sequestrum. In the end, what remaining iliac bone hadn't resorbed was subsequently resected. The decision of a partial resection leaving the outer table is, in hindsight, not to be recommended. The failure of the technique may be explained by the occurrence, immediately after the operation, of an extensive haematoma on the inner and outer fossae of the remaining iliac wing, which prevented the adhesion of the periostum and the iliacus and gluteus muscles. Deprived of vascularization by the periosteum, the remaining iliac wing necrosed and produced a sequestrum. However, resection of the inner iliac wing without exposing the outer part was technically challenging. Without a good view of the outer part it was difficult to evaluate the direction of progression of resection.

Rules of surgical resection of an aggressive malignant tumour of the pelvis could have been used. This tumour would have been

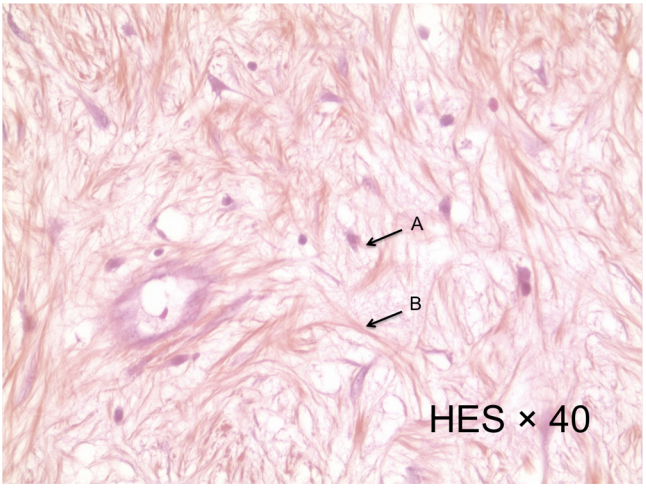

Fig. 5. Photograph showing the microscopic findings within the tumour. The tumour cells are fibroblasts with no nuclear atypia (A). The extracellular matrix is composed of collagen fibres (B).

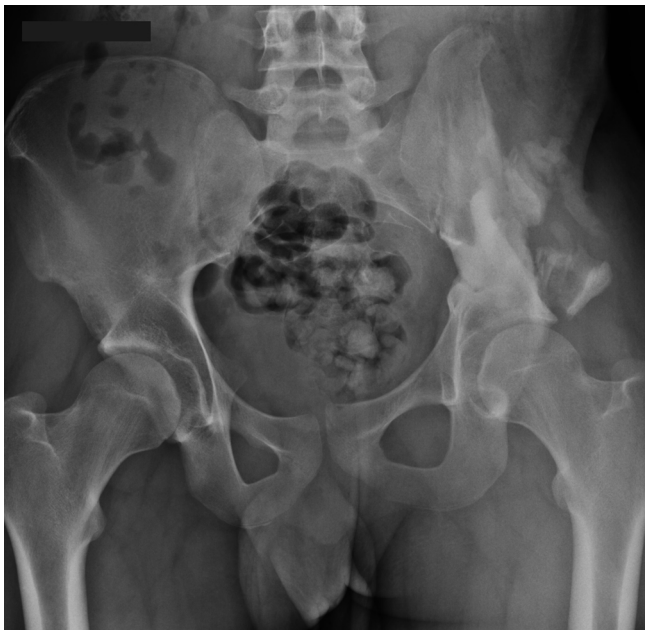

Fig. 6. AP view radiograph of the pelvis at last follow-up showing the lack of left iliac wing after resorption and resection of the sequestrum.

in Enneking's zone 1[18,19] and a complete resection of the iliac wing would have been recommended with no associated reconstruction. Indeed, for malignant bone tumours in Enneking's zone 1 with no fracture of the pelvic ring, Puget[20] proposed a simple resection. Reconstruction is necessary only when the pelvic ring is interrupted, the resection of the iliac wing being extended to the sacrum[20].

The most important complications encountered in the surgery of pelvic tumours are due to the length of the operation and heavy bleeding during and after the operation. Vascular wounds may occur. Dependant on the site and the type of pelvic tumours, the rate of repeated surgery because of post-operative complications is over 50%. Infections or mechanical problems may also occur when reconstructive surgery with insertion of prosthetic implants is necessary[20]. Functional results are related to the necessary sacrifice, in the case of malignant tumours, of the gluteus with more or less severe limping, which may necessitate the use of crutches. In spite of the important risk of weakening the gluteus medius, it appears, in hindsight, that complete resection of the iliac wing would have been easier and would have caused fewer complications than partial resection, which in our case yielded the same functional results.

The therapeutic procedure when dealing with a desmoplastic bone fibroma, particularly of the pelvis, is based on the limited experience of each surgeon, considering the rarity of this tumour[13].

In this case, the authors have attempted a partial resection of the iliac wing but unfortunately this theoretically appealing technique did not bring the expected results in this case. Therefore, the authors advise a simple resection of the iliac wing for this type of tumour in this location.

Conflict of Interest

None

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None

Ethical approval

Written informed consent was obtained

Author contributions

R.C. Rouchy and A. Courvoisier, study design and writing; S. Wimsey, data analysis and english writing; E. Bourgeois, data analysis and writing; B. Burroni, histological data analysis; J. Griffet, data analysis.

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