Case Report

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"Quasimodo" maybe had a giant liposarcoma of the back!

Cyrine Marmech[1]; Fouzia Hali[1]; Kenza Baline.[1]; Farida Mernissi[2]; Soumya Chiheb[1]; Abdeljabbar Messoudi[3]; Sidi Elhassen[3]; Abdelhak Garch[3].

1 Department of Dermatology and Venereology.
2 Department of Pathological Anatomy.
3 Department of Orthopedics and Traumatology.
Ibn Rochd University Hospital, Casablanca, Morocco.

Corresponding author: **Cyrine MARMECH**

MD, Ibn Rochd University Hospital, Department of Dermatology and Venerology, 1, Hospitals district, Casablanca;
E-mail address: cyrine.marmech1992@gmail.com

Abstract

Well-differentiated liposarcoma is called atypical lipomatous tumor when located in the extremities or in the trunk to differentiate them from their counterparts arising in the retroperitoneum or mediastinum. The atypical lipomatous tumors are rarely located on the back. We report a case of giant atypical lipomatous tumor of the back in 70-year-old patient presented with a huge mass of the back. A surgical biopsy revealed atypical cells with positive MDM2 testing at immunohistochemistry. Then a magnetic resonance imaging was realized and suggested the diagnosis of a slightly reshaped lipoma. Complete en-bloc tumor excision was performed. The specimen measuring 29x25x18 cm and weighs 5 kg 570g. Microscopic examination revealed differentiated mature adipocytes. Immunohistochemistry showed the expression of MDM2, indicating an atypical lipomatous tumor.

Resumé

Le liposarcome bien différencié est appelé tumeur lipomateuse atypique quand il est localisé aux membres et au dos pour le différencier du liposarcome bien différencié de localisation médiastinale et rétropéritonéale. La tumeur lipomateuse atypique est rarement localisée au niveau du dos. Nous rapportons un cas de tumeur lipomateuse atypique géante du dos. Il s'agit d'un homme de 70 ans qui a consulté pour une énorme masse du dos. La biopsie a montré des cellules atypiques avec expression duMDM2. L'IRM était en faveur d'un lipome légèrement remanié. Une exérèse complète en monobloc de la tumeur a été réalisée. La pièce d'exérèse pesait 5kg 570g et mesurait 29x25x18cm. L'étude microscopique montrait une prolifération d'adipocytes avec expression du MDM2 concluant à une tumeur lipomateuse atypique.

Key words: Atypical lipomatous tumor, Well-differentiated liposarcoma, Giant liposarcoma, Immunohistochemistry, Lipoma, Liposarcoma of the back.

Mots clés : Tumeur lipomateuse atypique, Liposarcome bien différencié, Liposarcome géant, Immunohistochimie, Lipome, Liposarcome du dos.

I- Introduction

Soft tissue sarcomas are mixed group of neoplasia that include more than 50 histological subtypes with various biological behaviors [1]. Liposarcoma is the most common subtype known as a malignant tumor of mesenchymal origin [2]. However, due to delayed diagnosis and treatment, it can reach a significant size and weight. The location of a liposarcoma in the back is extremely rare. We report a case of giant atypical lipomatous tumor/well-differentiated liposarcoma (ALT/WDLPS) of the back.

II- Case report

A 70-year-old male patient, presented with a painless mass at the back evolving for 30 years, for which he underwent surgical excision 4 times. The last one done 19 years ago with recurrence of the mass since 10 years. Physical examination revealed a large mass on the back measuring 29x25x18cm (Fig 1).

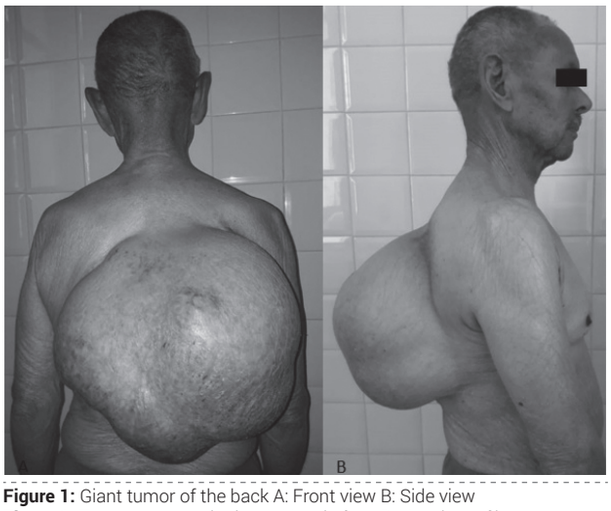

Figure 1: Giant tumor of the back A: Front view B: Side view
Figure 1: Tumeur géante du dos A: Vue de face B: Vue de profil

On palpation it was soft and mobile with clear and multilobed borders. The surgical biopsy revealed atypical cells with a hyperchromatic nucleus and positive MDM2 at immunohistochemistry. The MRI showed a large dorsal superficial soft tissue mass with a spontaneously hyperdense appearance in T1 and T2 and which fades on the fat saturation sequence, which confirms its fatty content. This mass is slightly reworked and does not enhance after injection of gadolinium, with no abnormalities of the opposing bone structures. The appearance of MRI suggested the diagnosis of a slightly reshaped lipoma. The thoraco-abdomino-pelvic CT scan was unremarkable. After a multidisciplinary meeting. The patient underwent a complete, en-bloc excision of the tumor. The pathological examination of the specimen showed a polylobate tumor measuring 29x25x18 cm and weighs 5 kg 570g (Fig 2). Microscopic examination revealed a proliferation of differentiated mature adipocyte with no atypia, arranged in lobules with positive expression of MDM2 at immunohistochemistry. The completely resected lipomatous mass has a healthy margin of 1mm .The final histologic diagnosis was an ALT. The post operative course was uneventful. No recurrence was reported after two years of follow-up (Fig 3).

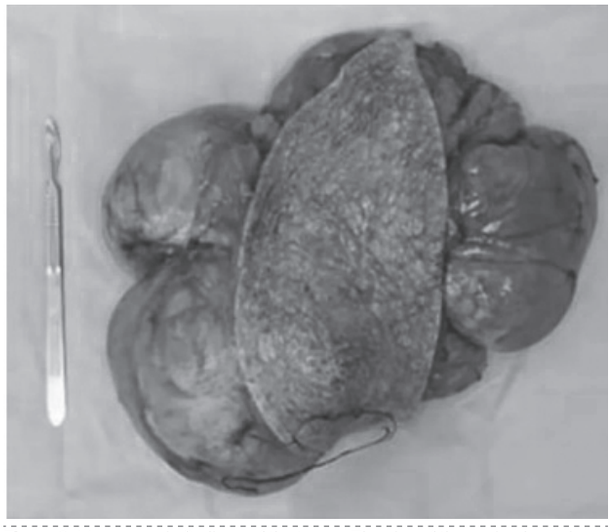

Figure 2: Macroscopic findings of the tumor. The size of the excised specimen is 29x25x18 cm
Figure 2: Aspect macroscopique de la tumeur. La taille du spécimen excisé est de 29x25x18 cm

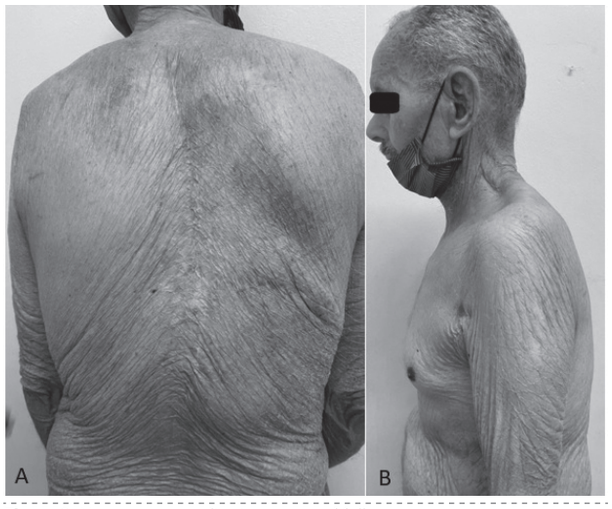

Figure 3: No recurrence after two years of follow-up.
Figure 3: Pas de récidive à 2 ans de recul

III- Discussion

According to the World Health Organization soft tissue classification, four major subtypes of liposarcoma are recognized: ALT/WDLPS, dedifferentiated liposarcoma (DDLPS), myxoid liposarcoma and pleomorphic liposarcoma [3]. ALT/WDLPS is a painless, locally invasive, non-metastatic tumor [4]. Its incidence peaks in the sixth and seventh decades of life. However, few cases were reportedat an earlier age, as for our patient.ALT/WDLPS represents 15% of primary soft tissue sarcomas of the chest wall. It is less frequently found around the head and neck, rarely on the back as seen in our case [5].

The ALT in this case is one of the biggest tumors reported in the literature. In the study of Klimstra the tumoral sizes ranged from 6 to 40 cm with a mean weight of 1kg500g [6]. In our observation, the weight was 5kg570g, which considered historical.

It is noted that the largest fatty tumors are liposarcomas. However, lipomas are usually small in size but can also

become giant [7,8]. The differential diagnosis depends on histopathological evaluation that allows the assessment of mitotic activity, cell atypia, necrosis and invasion, particularly due to the fact that distinguishing between liposarcoma and lipoma based on imaging is difficult [9,10]. Contrary to lipoma, ALT/WDLPS is histologically described by atypical stromal cells and lipoblasts in mature fat, usually with prominent sclerotic components [11,12].

In case of absence of complete histological findings, immunohistochemical staining for MDM2 and CDK4 can be performed to identify ALT/WDLPS [13-15].

Another diagnostic difficulty is differentiating between ALT/WDLPS and DDLPS, which is a biphasic tumor composed of WDLPS and non-fatty derived sarcoma. MDM2 and CDK4 can be over expressed in both [2]. In our case, the diagnosis of ALT was based on correlation of clinical, radiological and histological findings. The anatomic constraints in some circumstances often make it difficult to achieve negative margin surgical resection at the microscopic level. This unfortunate reality is the source of the high local recurrence rate seen in WDLPS tumors [1]. The ability to completely resection remains the most important predictor of local recurrence [4]. The ALT are non-radiosensitive tumors with low metastatic potential [16]. Surgical resection is the main therapeutic option. Our patient was treated only by complete surgical excision with no recurrence after 2 years of follow-up.

IV- Conclusion

ALT/WDLPS represent the most common subgroup of liposarcomas. It is locally aggressive and appears as a slow-growth, asymptomatic tumor. Only histologic and molecular analysis can provide an accurate diagnosis. Complete excision of the tumor remains the main treatment option. Our observation is distinguished by the historical weight as well as the unusual localization of the tumor.

V- Conflict of interest:

None

VI- References

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