Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Radiology

Desmoplastic Fibroma of the Mandible: Report of a Case

Zahira Benzenzoum[1*], Koussay Hattab[1], Nadia Mansouri[1], Aziz Zakaria[1], Mohamed El Bouihi[1], Saad Fawzi[1]

[1]The University Hospital Centre Mohamed VI of Marrakesh, Morocco

DOI: 10.36347/sjmcr.2023.v11i02.004 | **Received:** 14.12.2022 | **Accepted:** 25.01.2023 | **Published:** 05.02.2023

***Corresponding author:** Zahira Benzenzoum
The University Hospital Centre Mohamed VI of Marrakesh, Morocco

Abstract **Case Report**

Introduction: Desmoplastic fibroma is a benign and rare intraosseous tumor. This lesion has local aggressiveness and a significant risk of recurrence. It can simulate a malignant tumor chart. ***Observation:*** A 11-year-old child who consulted for a mandibular swelling gradually evolving for 1 year. Swelling is associated with dental mobility without sensory disorders or cervical lymphadenopathy. Radiological signs are in favor with aggressive pathology. The biopsy revealed a desmoplastic fibroid or low-grade fibrosarcoma. Radical surgical treatment was performed. ***Discussion:*** This location is exceptional. No cases of mandibular localization have been reported in the literature. The clinical and radiological signs are not very specific. The diagnosis is most often pathological associated with histochemical immune. The recommended management is surgical by a wide resection.

Keywords: Fibroid, desmoplastic, mandible, surgery.

INTRODUCTION

Desmoplastic fibroid, or bone desmoid fibroid, is a rare benign bone tumor of fibroblastic nature [1, 2]. The initial description of this tumor goes back to Jaffe [3] in 1958. There is histological and macroscopic similarity between desmoplastic fibroids and soft tissue desmoid fibroids [4, 5].

The definition of the World Health Organization (WHO) [6] is: "A benign but locally aggressive bone tumor that is characterized by the abundant formation of collagen fibers by tumor cells"; the tumor tissue is "poorly cellular and the nuclei are ovoid or elongated in shape"; "The cellularity, pleomorphism and mitotic activity of fibrosarcoma are absent." A case of desmoplastic fibroma supported in our department is reported here to identify specific diagnostic and therapeutic problems that may arise during the management of this rare tumor.

OBSERVATION

A 12-year-old child consulted in our training for a right mandibular swelling of progressive evolution over a period of 1 year, at the clinical examination without neurological or noticeable signs.

The exooral clinical examination (Figure 1) found facial asymmetry related to a swelling forming part with the painless right mandible, not adherent to the skin, fixed in relation to the deep plane, with healthy skin in appearance. Endooral examination (Figure 2) revealed a good oral opening, and a tumor of the lower right premolar region measuring 4 cm of major axis. The swelling filled the vestibule and extended from the floor. The mucosa was healthy and without dental mobility. On palpation, the lesion was painless and of firm consistency. There was no sensory disturbance in the territory of the chin nerve. Examination of the cervical lymph node areas did not show lymphadenopathy.

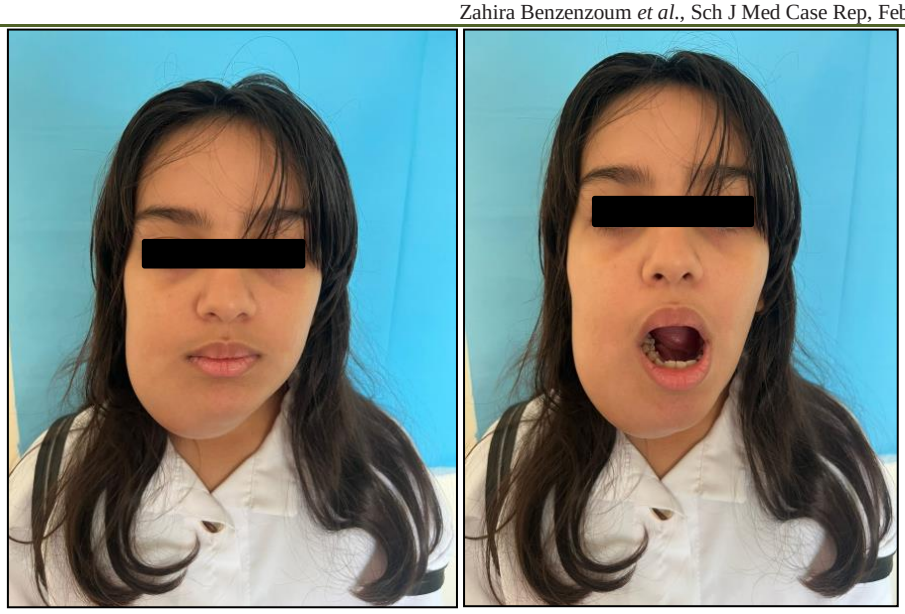

Fig. 1 & 2: Exo and endooral view showing facial asymmetry at the expense of a right mandibular swelling with a good oral opening

An X-ray performed, made of an orthopantomogram (Figure 3) and a facial CT scan (Figure 4) with axial, coronal and 3D sections found a well-limited right mandibular osteolytic lesion of regular contours measuring 56X47X49 cm centered on the horizontal branch, the angle and part of the rising branch. And at the panoramic radio a polylobed osteolytic lesion of 7.5 cm of the right hemimandibule mainly affecting the right horizontal branch, the mandibular angle and the homolateral rising branch.

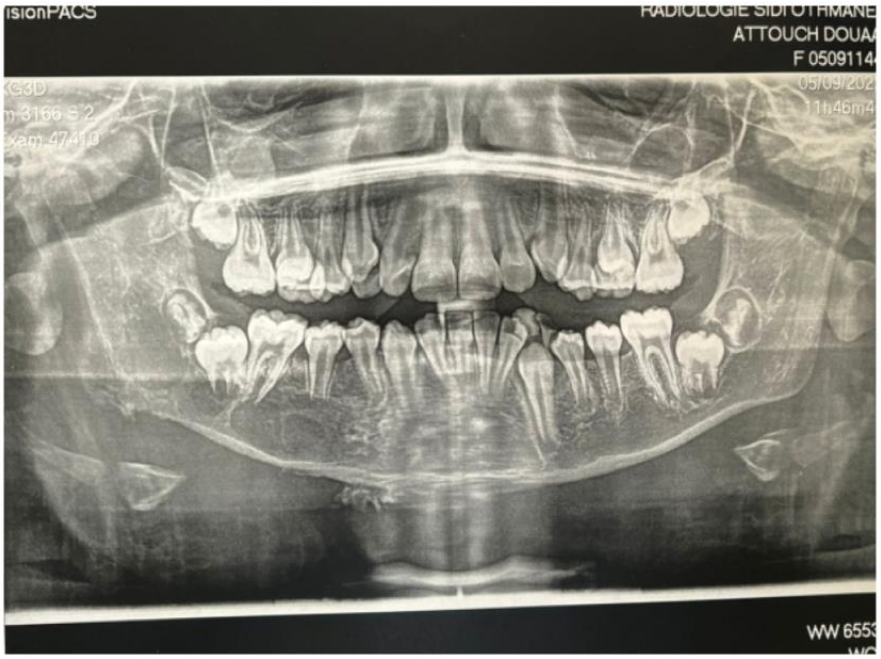

Fig. 3: Image 7.5 cm polylobed osteolytic lesion of the right hemimandibule mainly affecting the right horizontal branch, the mandibular angle and the homolateral rising branch

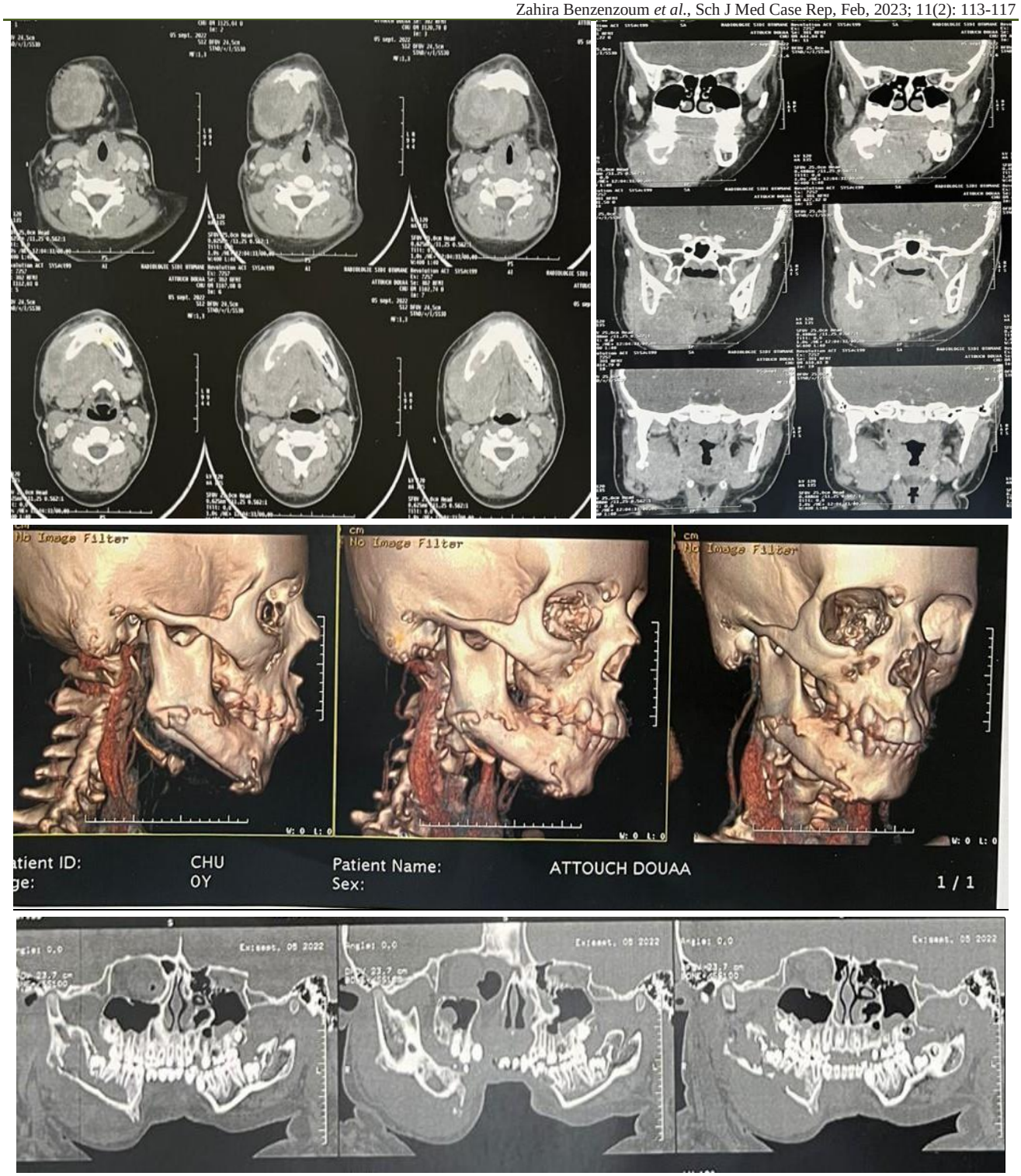

Figure 4: Facial scanner with axial, coronal and 3D sections objectifying a well-limited right mandibular osteolytic lesion of regular contours centered on the horizontal branch, the angle and part of the rising branch

A surgical bone biopsy was performed by a direct approach. The histological diagnosis was that of desmoplastic fibroid or low-grade fibrosarcoma.

A right hemimandibulectomy (Figure 5) was performed under general anesthesia by transfacial approach in our patient carrying the inferior alveolar nerve. An immediate reconstruction by a maxi plate was carried out. The histopathological examination of the operating room revealed a desmoplastic fibroid measuring 9cm. The postoperative follow-up was simple. On a general level, a consultation with a child psychiatrist took place in order to improve the quality of life of the little one in front of the mutilating surgery she took place.

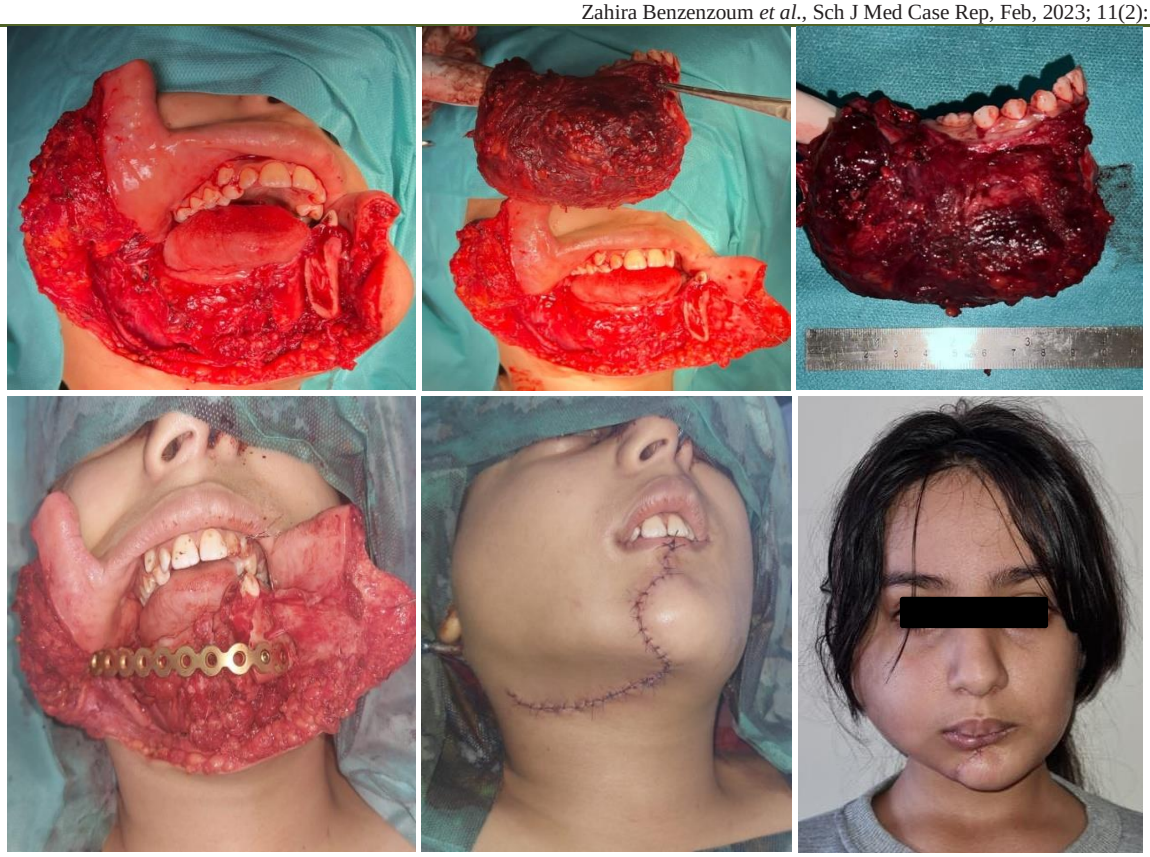

Figure 5: right switch hemimandibulectomy with maxi plate reconstruction in our patient with post-operative view (3 months after the gesture)

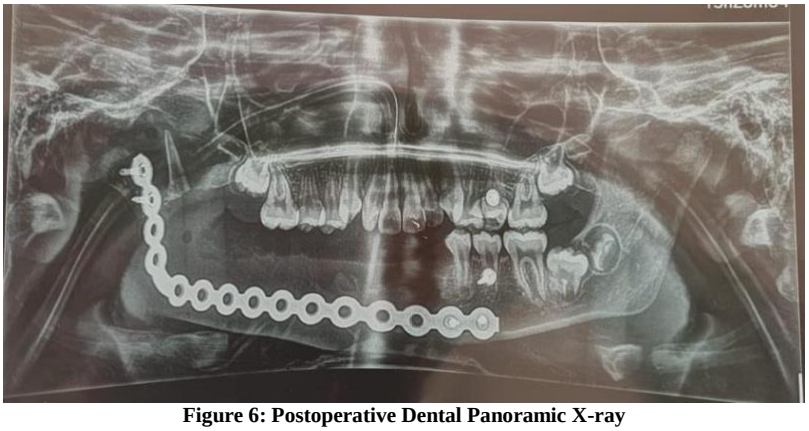

Figure 6: Postoperative Dental Panoramic X-ray

DISCUSSION

Desmoplastic fibroids appear as a very rare bone tumor. It represents, according to Taconis *et al.*, [7] 0.3% of benign bone tumors. There is no sexual predominance and desmoplastic fibroids occur in more than 75% of cases in the first three decades. Clinical signs are variable is often discreet. In most cases, it is a little or no painful tumor. Slow growth may explain some delay in management, scannographically, the lesion has areas of uni or multifocal osteolysis with or without peritumoral osteocondensastion border. The intralesional partitioned appearance is described as a honeycomb or honeycake structure. Invasion of the soft parts and rupture of the cortical are found in 20% of cases [9]. The differential diagnosis raised by imaging is: giant cell lesion, intraosseous angioma, eosinophilic granuloma, aneurysmal cyst, chondromyxoid fibroid, ossifying fibroid, ameloblastoma, sarcomas and metastases.

This finding helps to explain, as is the case in our observation, the extremely high recurrence rate in cases where intralesional excision of the desmoid fibroid was performed, either by simple enucleation or by simple curettage. This relationship between the tumor tissue and the adjacent bone justifies extra-lesional excision carrying a margin of healthy peritumoral bone tissue to limit the risk of local recurrence. Anatomopathologically, there is a great histological similarity of bone desmoid fibroid with soft tissue desmoid fibroids. It is a connective tissue very

rich in collagen. These collagen fibers are arranged in large parallel bundles or on the contrary in fine corrugated fibers without clear fasciculate arrangement. Cell wealth is moderate or even low. The cells are fusiform and small. The nuclei are small, regular, round or ovalar, with fine chromatin. Mitoses are rare. There is no mitotic atypia. Desmoplastic fibroids have moderate vascularization. There are no bone or cartilage components within the tumor. The most delicate histological differential diagnosis is low-grade fibrosarcoma [7, 10].

Tumor biopsy is an essential therapeutic prerequisite. It should preferably be surgical in order to provide sufficient material to the pathologist. In our observations, the initial surgical biopsy made it possible to establish the diagnosis in our case. The natural history of desmoplastic fibroids after treatment is marked by the high risk of tumor recurrence. The tumor recurrence finds a tumor with the same histological features as the initial tumor. In contrast, described cases of malignant transformation into fibrosarcoma are most often considered to be underestimates of the exact nature of the initial tumor. The same applies to the occurrence of metastases. Desmoplastic fibroma is a benign tumor, with local progressive malignancy, not giving metastasis or malignant transformation [8, 11].

Analysis of desmoplastic fibroid treatment in the literature is difficult. Indeed, the therapeutic conduct is based on the necessarily limited experience of each author, given the rarity of this tumor. Nevertheless, it is very clear that intra-lesional procedures, such as curettage, are doomed to a very high recurrence rate, from 50 to 72% [8]. Extralesional resection procedures offer the best outcome, with less than 5% recurrence [8]. Desmoplastic fibroid, however, remains a benign tumor and, depending on the location, an extra-lesional resection procedure of the tumor may be functional prognosis. Reconstruction after resection uses the usual techniques of tumor surgery: vascularized autograft or not, allograft.

Postoperative monitoring should be prolonged. Indeed, the average time of occurrence of recurrences is three years, it must be clinical and radiological [8].

CONCLUSION

Desmoplastic fibroma (bone desmoid fibroid) is a rare bone tumour. The histology of desmoplastic fibroma is identical to that of soft tissue desmoid fibroids. It is a slow-growing tumour. For radiological diagnosis, CT or magnetic resonance imaging (MRI) would be an examination of choice. It makes it possible to take stock of the local extension, at the level of the bone and possibly the soft parts. A biopsy is an essential prerequisite for treatment. The positive diagnosis is histological. Differential histological diagnosis is sometimes difficult, especially with low-grade fibrosarcomas. The evolution after treatment is marked by a high rate of local recurrence in case of incomplete excision (curettage). The treatment of choice is surgical and consists of a large tumor resection whenever tumor localization allows [12].

REFERENCES

1. Anract, P., De Pinieux, G., & Tomeno, B. (2001). Fibrome desmoïde intra osseux. In: Encycl Méd Chir, Appareil Locomoteur, Elsevier, Paris, p. 14-752.
2. Danan, J. P. (1974). Fibromes desmoïdes de l'os. In: Tumeurs osseuses bénignes, Monographies Annales Chirurgie, Expansion Scientifique Francaise, p. 60-64
3. Jaffe, H. L. (1958). Desmoplastic fibroma and fibrosarcoma. In: Tumors and tumorous conditions of the bones and joints. Lea and Febiger, Philadelphia, p. 298-303.
4. Meary, R., Danan, J. P., Tomeno, B., Forest, M., & Cirotteau, Y. (1978). Fibromes desmoïdes des membres. *Rev Chir Orthop*, 64, 195-204.
5. CHATELARD, P. A., Gilly, F. N., Carret, J. P., Vauzelle, J. L., Brunat, M., & Braillon, G. (1991). Tumeurs desmoïdes extra-abdominales: indications thérapeutiques: à propos de 28 cas. *Acta orthopaedica belgica*, *57*(3), 227-233.
6. Schajowicz, F. (1981). Other connective tumors, desmoplastic fibroma. In: Tumors and tumorlike lesions of bone and joints. Springer-Verlag, New York, Heidelberg, Berlin, p. 335-339.
7. Taconis, W. K., Schutte, H. E., & Van Der Heul, R. O. (1994). Desmoplastic fibroma of bone: a report of 18 cases. *Skeletal Radiol*, 23, 283-288.
8. Bohm, P., Krober, S., Greschniok, A., Laniado, M., & Kaiserling, E. (1996). Desmoplastic fibroma of the bone. A report of two patients, review of the literature, and therapeutic implications. *Cancer*, 78, 1011-1023.
9. Crim, J. R., Gold, R. H., Mirra, J. M., Eckardt, J. J., & Bassett, L. W. (1989). Desmoplastic fibroma of bone: radiographic analysis. *Radiology*, *172*(3), 827-832.
10. Dahlin, D. (1978). Fibrosarcoma and desmoplastic fibroma. In: Bone tumors, general aspects and data on 6,221 cases. Charles C. Thomas, Springfield, p. 315-328.
11. Vanhoenacker, F. M., Hauben, E., De Beuckeleer, L. H., Willemen, D., Van Marck, E., & De Schepper, A. M. (2000). Desmoplastic fibroma of bone: MRI features. *Skeletal radiology*, *29*(3), 171-175.
12. Puget, J., & Utheza, G. (1986). Reconstruction de l'os iliaque avec l'aide du fémur homolatéral après résection pour tumeur pelvienne. *Rev Chir Orthop*, 72, 151-155.