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MALIGNANT SOLITARY FIBROUS TUMOR: A REPORT OF TWO CASES

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ABSTRACT

Case Report 1: A 39-year-old woman presented with a tumefaction of the left thigh, measuring 13 centimeters in diameter. A chest, abdomen and pelvis computed tomography (C.A.P. CT) showed a mass measuring 20 cm, located in the left psoas muscle, with extension to the pelvis, and pulmonary metastases. Histopathological analysis of the specimen was consistent with malignant solitary fibrous tumor.

Case Report 2: A 80-year-old man presented with a mass of the right nasal cavity. Cranio-facial MRI showed the presence of a tumor of the right nasal cavity with intracranial extension. Histopathological analysis of the specimen was consistent with malignant solitary fibrous tumor.

The solitary fibrous tumor is a rare mesenchymal tumor. It is preferentially located at pleura. Extra-thoracic localization is rare. In soft tissue, solitary fibrous tumor accounts for approximately 0.6% of tumors. The sino-nasal localization is very rare. Histologically, the tumor is made of a dense proliferation of fusiform cells. The immune-histochemical study of solitary fibrous tumor classically shows a diffuse expression of vimentin and CD34. Intense and diffuse nuclear staining of STAT-6 is highly characteristic of this tumor. The treatment of choice is the complete surgical resection.

Keywords: Malignant; Solitary fibrous tumor; Nasal cavity; Soft tissue.

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INTRODUCTION

Malignant solitary fibrous tumor (MSFT) is a rare mesenchymal tumor. The classic criteria for diagnosis of malignancy are high cellularity, high mitotic activity, polymorphism, haemorrhage and necrosis. It is preferentially located in pleura; however it is not restricted to the serous surfaces. Various localizations have been reported in the literature. Among these exceptional locations, we present two observations of a malignant solitary fibrous tumor of the soft tissues of the thigh and of the nasal cavity. The diagnosis was confirmed by histological and immune-histochemical analysis.

CASE REPORT 1

A 39-year-old woman presented with a tumefaction of the antero-internal part of the left thigh. The clinical examination found a poorly limited mass, measuring 13 centimeters in diameter, not painful and without inflammatory signs.

A C.A.P. CT showed a mass measuring 20 cm, located in the left psoas muscle, with extension to the pelvis, and pulmonary metastasis. The bone scintigraphy was normal. A tumor biopsy was performed. Histological examination revealed an heterogeneous cellularity (**Fig. 1, 2**).

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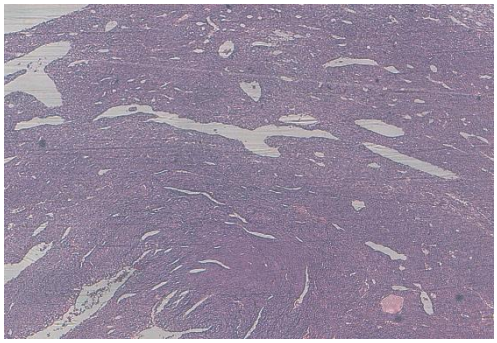

Figure 1: Fusocellular tumor proliferation (HESx10)

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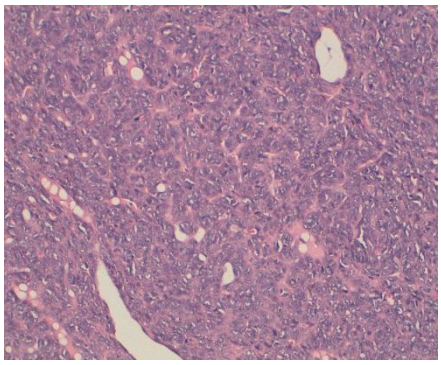

Figure 2: tumor proliferation made of fusiform cells (HESx20)

Tumor cells were spindle and presented cytonuclearatypias with numerous mitosis (12 mitosis/ 10 high-power fields) (Fig. 3).

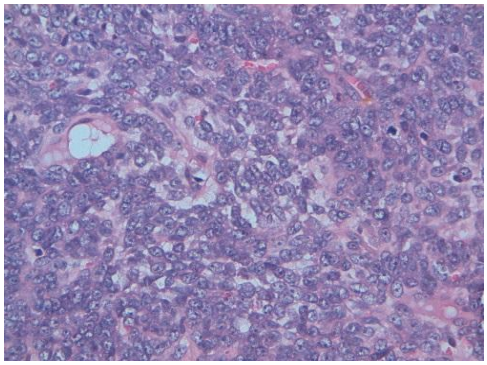

Figure 3: Tumor cells show obvious cytonuclear atypia with multiple mitotic figures (HESx40)

Immuno- histochemical study showed a positivity of the anti-CD34 and the anti-BCL2 antibodies (**Figures 4, 5**).

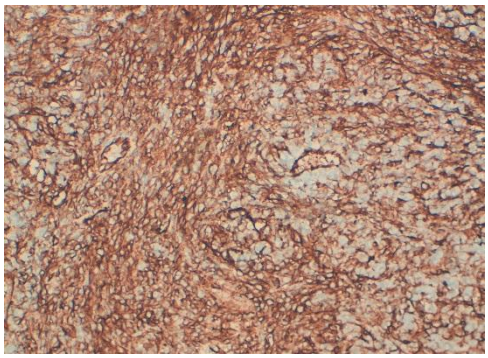

Figure 4: Diffuse positivity for CD34

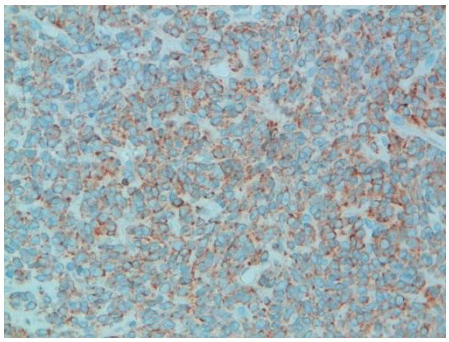

Figure 5: Bcl-2 positivity

The diagnosis of malignant solitary fibrous tumor was retained and the patient received six cures of chemotherapy and died before the end of cures.

CASE REPORT 2

Eighty year-old man presented with a process in the right nasal cavity extended to the anterior cerebral level. The clinical examination found a complete obstruction of the right nasal cavity by a tumoral mass. Cranio-facial MRI showed the presence of a process of the right nasal cavity with intracranial extension. An endoscopic excision of the tumor was performed.

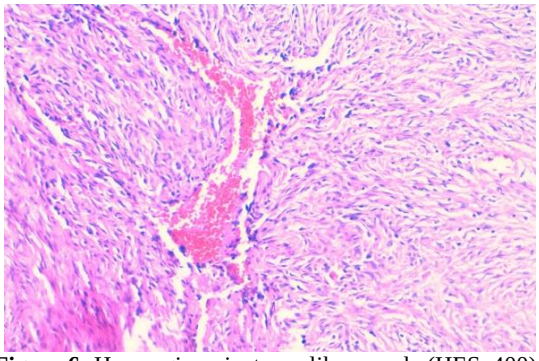

Figure 6: Hemangiopericytoma-like vessels (HESx400).

Histological examination revealed high cellular proliferation made of bundles and storiform structures. Tumor cells were fusiform with moderate cytonuclear atypias. There were 6 mitosis/ 10 high power fields. Hemangiopericytoma-like vessels were noted (**Figure 6**). This proliferation infiltrated the bone tissue (**Figure 7**) and came in contact with the cerebral parenchyma. Immuno-histochemical studies revealed a strong positivity for CD34 (**Figure 8**) and STAT6 with a proliferation index Ki67=20%.

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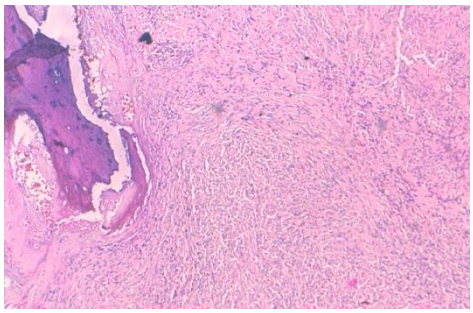

Figure 7: Tumoral proliferation infiltrating the bone tissue (HESx200).

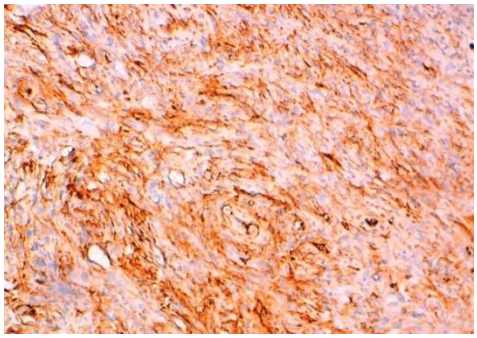

Figure 8: Immuno-histochemical studies revealed a strong positivity for CD34

The diagnosis of malignant solitary fibrous tumor was retained. The patient received post-operative radiotherapy. A cerebral scan performed one month after the surgical excision did not show any signs of tumor recurrence.

DISCUSSION

The solitary fibrous tumor (SFT) is a rare mesenchymal tumor. The first description was made by Klemperer and Rabin in 1931 [1]. It is developed preferentially at the pleural level. Recently, multiple extra- thoracic sites have been reported [2-3].

In soft tissue, SFT accounts for approximately 0.6% of soft tissue tumors [4]. The sino-nasal localization is rare, as is the intracranial extension. Only 22 cases have been reported in the literature [5].

This tumor occurs equally without distinction of sex [1]; it is observed in all age groups.

The low clinical aggression, the ubiquitous nature and the ability to simulate a multitude of neoplasms make the diagnosis of these mesenchymal tumors difficult [6].

Currently, according to several histological and immunohistochemical studies, it seems clear that these tumors are non mesothelial mesenchymal tumors of (myo) fibroblastic origin [7-8] and are comparable to those found in extra- thoracic location [9].

Clinically, the majority of extra- thoracic TFS are asymptomatic. After a long evolution, the appearance of a painful mass remains the main sign [2]. It manifests as a mass of variable size, but of regular limits [10] and most often mobile. These tumors can radiologically mimic any mesenchymal lesion. The final diagnosis remains pathological. Macroscopically, the tumor is well limited and often encapsulated and translucent. The size varies generally between 1 cm and 25 cm [10]. At the cut, the surface appears fasciculate and lobulated. Its color is often greyish, sometimes pink-white [10].

Histologically, the tumor is made of a dense proliferation of fusiform cells, dispersed orderly and supported by a frame of collagen fibers of variable abundance. It is characterized by branching hemangiopericytoma- like vessels. The main signs of malignancy are represented by a high mitotic index (greater than four mitoses per ten fields at high magnification), a high cell density, necrotico-haemorrhagic changes, a marked nuclear pleomorphism and vascular invasion [11 -12-13-14]. In both presented cases, the criteria for malignancy found are the high cell density, the cytonuclear atypias and the high mitotic activity.

The immune- histochemical study of SFT classically shows a diffuse expression of vimentin and CD34, a variable expression of CD99 and bcl-2 protein and negativity of the epithelial markers (cytokeratin, EMA) and of the protein S100 [15]. Intense and diffuse nuclear staining of STAT-6 is highly characteristic of SFT, seen in more than 90% of the cases [16].

Some factors of poor prognosis have been reported as the loss of expression of the CD34 antigen, the high expression of Ki67 (MiB1) and of the P53 protein [17].

Microscopically, there are several differential diagnoses such as: fibrous histiocytoma, fibromatosis, fibrosarcoma and hemangiopericytoma. Immuno- histochemical study helps in distinguishing these entities [10].

The treatment of reference is the complete surgical resection. Incomplete resection is a pejorative factor of ulterior evolution [11, 17]. The role of other treatments is not yet well codified. Chemotherapy may be interesting in inoperable malignant forms, in neo-adjuvant, in large tumors, or in adjuvant, in cases of incomplete resection [18].

Radiotherapy could also be indicated after surgical resection, in cases of histological signs of malignancy, especially if this resection is incomplete. Its real interest remains to be demonstrated.

The majority of recurrences occur within the first two years after surgery [14, 15]. Metastases have also been reported [19].

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CONCLUSION

MSFT is a rare tumor. Its diagnosis is histological, largely facilitated by immunohistochemistry. Surgery is the basis of treatment and the role of adjuvant therapies (chemotherapy and radiotherapy) remains to be clarified. Close control is necessary because of the risk of recurrence or metastasis.

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