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INTERNATIONAL JOURNAL OF ADVANCED RESEARCH (IJAR)

Article DOI: 10.21474/IJAR01/15442
DOI URL: http://dx.doi.org/10.21474/IJAR01/15442

RESEARCH ARTICLE

SOLITARY FIBROUS TUMOR OF THE SKIN: AN UNUSUAL LOCATION

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Manuscript Info

Manuscript History
Received: 25 July 2022
Final Accepted: 28 August 2022
Published: September 2022

Key words:-
Solitary Fibrous Tumor, Skin. Surgical Excision, Recurrences, Radiotherapy

Abstract

Background : Solitary fibrous tumours rare spindle cell neoplasms, initially regarded as occurring exclusively within the thoracic cavity with a pleural origin. We report an unusual and rare case, of solitary fibrous tumor of the skin. Whose evolution has been marked by several recurrences after surgical excision
Case Report: 49 year old man, with a dorsolumbar cutaneous tumor.

Introduction:-

Solitary fibrous tumor (SFT) is a rare spindle cell neoplasm originally described by Klemperer and Rabin in 1931 as a pleural based lesion. [1]

Due to this location, SFTs were thought to be of mesothelial origin. However, subsequent ultrastructural and immunohistochemical studies have shown the neoplastic cells to be fibroblastic or myofibroblastic in nature [2, 3]

Cases occurring outside the pleura have been reported in virtually all organs, including soft tissues, upper respiratory tract, ovary, orbit, liver, and retroperitoneum [4, 5, 6, 7].

We report an unusual case, because skin localization is extremely rare, and also because the evolution was marked by several recurrences.

Case Report:
A 49-year-old mediteranian man, with no particular medical history, presented with a dorso-lumbar cutaneous mass evolving since 06 months.

The examination found an oval brown mass, measuring 8 cm in diameter, firm, adherent but painless. (Figure 1)

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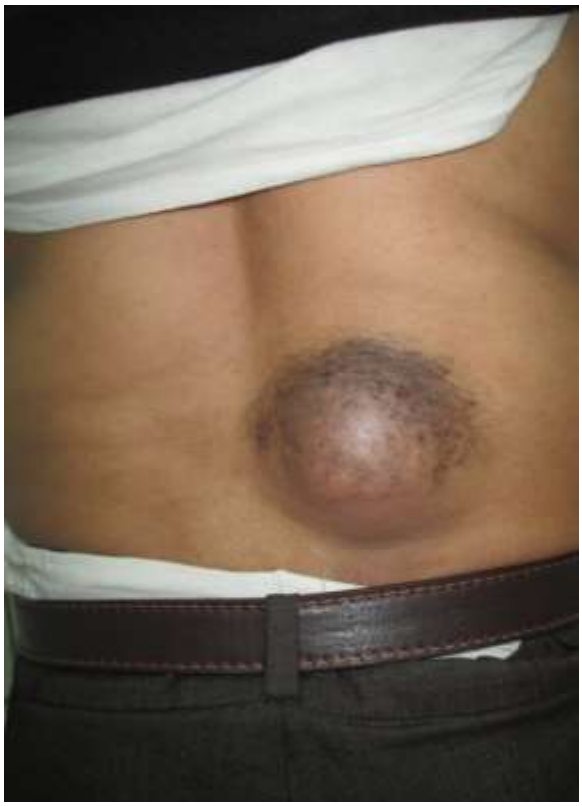

Figure 1:- Aspect of the tumor.

MRI showed the existence of an oblong formation of the superficial soft tissues, sitting on the right side of the lumbar spine, measuring 68 × 64 × 40 mm, well limited, heterogeneous containing areas of necrosis and septas, intensifying after injection of contrast product.

This process comes into contact with the left transversalis fascia without invading the fat or erector muscle of the lumbar spine (figure 2).

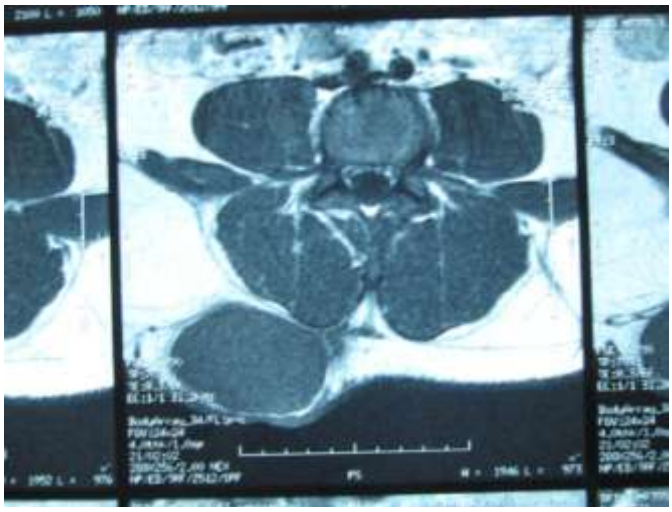

Figure 2:- MRI showing an oblong formation of the superficial soft tissue.

The tumor's excision was performed, and its histopathological nature was defined: it was a solitary fibrous tumor (figure 3)

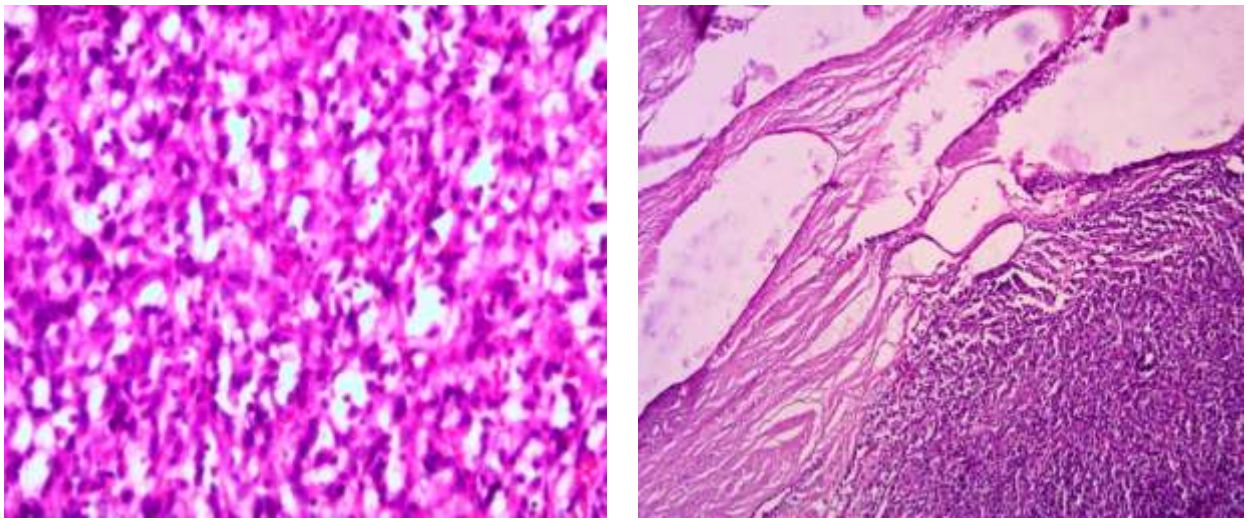

Figure 3:- Histopathological aspect of the solitary fibrous tumor: spindle cells.

The patient was lost, because he lived far from the hospital.

One year later, he presented with a local tumor recurrence, it sat at the surgical scar, it was a subcutaneous mass identical to the previous lesion: firm, painless and adherent to both superficial and deep plans (figure 4).

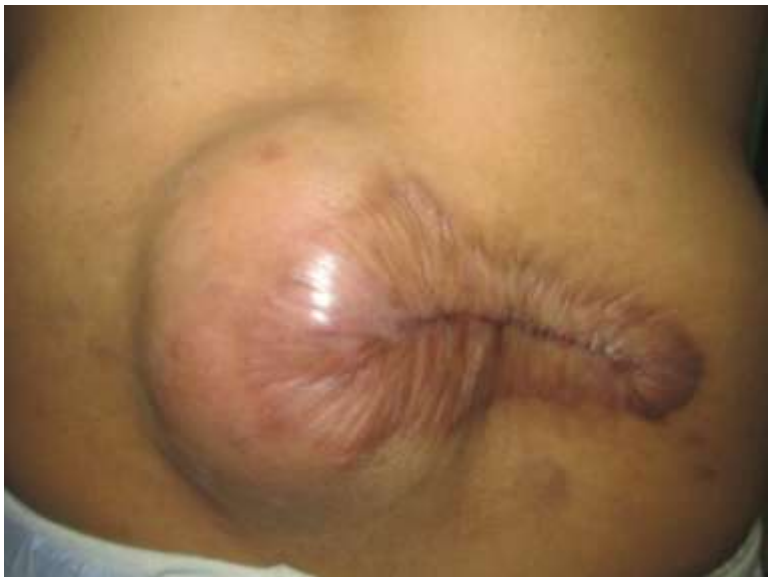

Figure 4:- First recurrence on surgery scar.

MRI shows the same pictures as before: a median lumbar tissue tumor, subcutaneous, oval, well limited, spreading from L2 to L5 and measuring 85mm height and 63mm wide. It was heterogeneous and intensely enhancing.

This mass infiltrated the overlying skin and respects the deep musculo-aponevrotic floor.

A surgical revision was decided and the histopathological exam shows a proliferation of spindle cells CD 34 +.

Six months later, the tumor has recurred again, the patient had a tumor excision for the third time, the wide wood loss was covered by a skin graft.

The surgery was followed by radiotherapy.

Unfortunatly, the tumor has recurred another time, and it was the last time, because the patient is dead four months after.

Discussion:-

Solitary fibrous tumors are rare tumors initially described in the pigs by Klemperer in 1931 as "localized fibrous mesotheliomas" [1].

In the literature, we find several terminologies for fibrous solitary tumors [8, 9] using in particular "mesothelioma", fibrous mesothelioma, localized mesothelioma, submesothelial fibroma, serous cavities' primary tumor, localized fibrous tumor.

Extrathoracic TFS accounts for approximately 0.6% of soft tissue tumors and occurs equally in both genders [8]

Clinically, it is a progressively larger, well-limited lesion occurring between 30 and 70 years of age [5, 10].

Some systemic signs (which disappear after tumor removal) have been described as arthralgia or osteoarthropathy as well as hypoglycaemia, which would be related to an insulin growth factor secretion (IGF) by the tumor [8, 10].

These tumors are characterized by their great morphological variability. Neverthless, two architectural profiles predominate:

1. The compact fusocellular appearance where the tumor cells are arranged according to a storiform architecture, fibrosarcomatous, neural or a little bit anyhow (patternless pattern)
2. the hemangiopéricytaire aspect where the tumor vessels appear anastomosed and typically branched in deer antlers.

At low magnification there is typically an alternation of hypercellular and hypocellular zones separated from each other by thick hyaline collagen keychains. These collagen bands are responsible for the macroscopic multinodular appearance of the tumor. Presence of stellate areas of dense collagen with peripheral palisades of nuclei. Myxoid rearrangements and areas of diffuse fibrosis are not uncommon.

At high magnification, fusiform cells have a scanty cytoplasm and wavy or circumflex nuclei as in neurofibromas or schwannomas. Chromatin is often faded, pale, and there may be intranuclear cytoplasmic inclusions. Epithelioid-like polygonal cells and / or multinucleate giant cells of osteoclastic type can also be observed.

The vascularization is marked hemangiopericy. Foci of hypercellularity can be observed in benign SFT as well as islets of mature adipose tissue. [9, 11, 13, 14, 15, 16]

Immunohistochemistry is important for the diagnosis of solitary fibrous tumors. CD34 labeling is strongly positive, showing diffuse cytoplasmic reactivity in 75-100% of cases, according to the studies, making it one of the most reliable markers. [13, 17, 18] . CD99 and Bcl2 are expressed in 50% of cases. Immunolabeling with beta-catenin and p53 has also been reported. [19, 20, 21, 22]

Some series report the focal expression of estrogen-progestogen receptors by tumor cells as a risk factor favoring recurrence after surgical excision. Smooth muscle actin is generally not expressed by tumor cells, despite the supposed (myo) fibroblastic histogenesis [23]. This contradiction is explained by the fact that there appear to be four different (myo) fibroblastic phenotypes, one of which does not express actin. [24].

20-30% of cases are variably positive for EMA and Bcl2. Focal reactivity to PS100, cytokeratin, and / or desmin has occasionally been reported. [15]

Regarding the clinical course, fibrous solitary tumors are usually benign tumors.For extrapleural tumors, recurrences and metastases seem rare. [17]

The risk factors for malignancy are invasive surgical margins, pejorative histological elements, a mitotic index greater than four mitoses per ten fields at 400-fold magnification, the presence of necrotic or hemorrhagic zones, a

cellular pleiomorphism and a high cell density. [10, 25, 26, 27, 28]. As well as a size larger than 10 cm, however, about 60% of benign cases have at least one of these criteria [9]

The p53, basic fibroblast growth factor (bFGF) and Ki67 markers would be more frequently positive in malignant cases, and the level of p53 would be correlated with the degree of malignancy which would make it a diagnostic marker but also a prognostic one. [29, 30]

The treatment of choice is the surgical excision which must be wide; however no consensus indicating the margins of excision of this type of tumor has been found in the literature. [5]

The solitary fibrous tumor remains a rare tumor, there is no real evaluation of adjuvant treatments [31, 32]

In this patient, considering the surgical difficulties, it was proposed a complementary irradiation. Radiological responses after radiotherapy for primary tumors or recurrence have also been reported. [33, 34]

Radiotherapy of solitary fibrous tumors has been rarely described, but its reported efficacy in cases of recurrence allows it to be used in cases where surgical excision alone would be considered insufficient

Regular and prolonged monitoring of these tumors is necessary [35]. In our case, we decided to do a monthly monitoring the first quarter, then every three months the first year and every six months the second year, then once a year. This surveillance is essentially clinical.

Conclusion:-

SFT extrathoracique is often aggressively malignant tumour with a poor prognosis, which is likely to recur even after a surgical treatment and radiotherapy.

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