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(CASE REPORT)

Thoracic schwannoma mimicking a Darier-Ferrand dermatofibrosarcoma: Diagnostic and reconstructive challenges

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Abstract

Primitive schwannoma refers to a rare benign tumor that originates from Schwann cells, which surround and insulate peripheral nerve fibers. These tumors are benign, exhibit slow growth, and have a very low risk of malignant transformation. They can remain asymptomatic for years or cause pain, paresthesia, or neurological deficits. When they develop in atypical regions, such as soft tissues or subcutaneous areas, they may be mistaken for other types of tumor masses.

This is the case in our study, which reports on a patient referred to us for a thoracic cutaneous tumor with a partial biopsy result suggesting a schwannoma. Treatment involved complete surgical excision. Histological analysis of the surgical specimen revealed a surprising finding: a dermatofibrosarcoma of Darrier and Ferrand (DFSP), often considered a differential diagnosis for schwannoma.

This study highlights both the importance of intraoperative frozen section analysis during surgery and the crucial role of the plastic surgeon in managing extensive and deep tissue defects that were unexpected based on the initial clinical diagnosis.

Keywords: Schwannoma; Dermatofibrosarcoma of Darrier and Ferrand; Thoracic; Latissimus dorsi flap

1. Introduction

Schwannoma and dermatofibrosarcoma of Darrier and Ferrand (DFSP) share certain similarities, despite originating from different cellular lineages. Both are mesenchymal tumors, typically slow-growing, and may present as painless cutaneous or subcutaneous nodules. Their diagnosis often relies on histopathological examinations, as their clinical presentation can be misleading, as observed in our patient. [1]

Additionally, both tumors have a potential for local recurrence, requiring meticulous surgical management with clear margins to minimize the risk of recurrence. However, their biological behavior differs: DFSP is more aggressive and carries a risk of malignant transformation, unlike schwannoma, which generally remains benign. [2]

Although surgery is the main treatment for both tumors, it should be adapted to their specific features for the best outcome.

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1.1. Patient observation

This is a 61-year-old male, a former farmer. He is a chronic smoker who quit 4 years ago, with no significant past medical history.

He was admitted for a plastic surgery consultation due to a left lateral thoracic mass that has been progressing for 16 years. The condition evolved with a gradual increase in size, the appearance of fistulas discharging cloudy fluid, and pain described as electric shocks, prompting the patient to consult a dermatologist 3 years ago. A biopsy performed at that time revealed an axillary schwannoma without signs of malignancy.

General Examination The patient is stable hemodynamically, respiratory-wise, and neurologically.

Pulmonary and Pleural Examination: Good thoracic expansion, a large fistulized mass in the anterior axillary line, and digital clubbing.

Cutaneous Examination Multiple subcutaneous masses are present, hard and fixed relative to the superficial plane but mobile relative to the deeper plane. The rest of the skin examination, both locally and at a distance, is unremarkable. (fig 1)

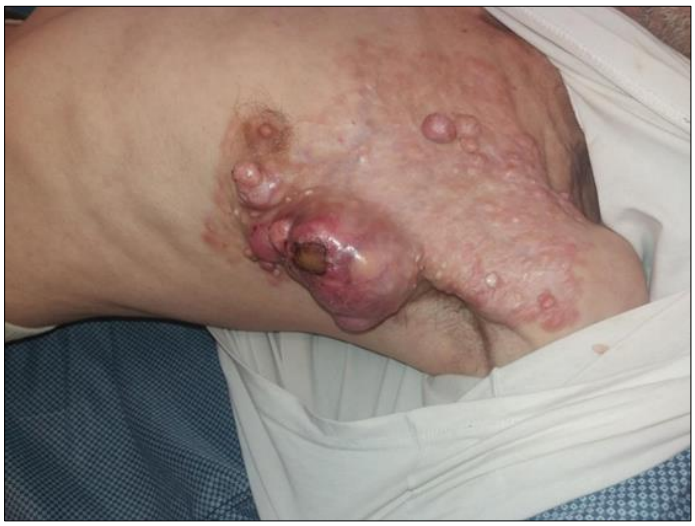

Figure 1 Aspect during the initial consultation

1.2. Other Clinical Examination: Normal findings.

A thoracic MRI was requested: presence of multiple lymphadenopathies in the left axillary region, the largest being necrotic and measuring 7.7 x 5.6 cm. Multiple bullae of emphysema in the upper lobes bilaterally, with bronchial syndrome. Skin thickening without muscular extension in the corresponding area. The abdominal MRI: unremarkable. Given the clinical, histological, and radiological findings, it was decided in consultation with the thoracic surgery department to proceed with: surgical excision with intraoperative frozen section analysis, left axillary lymph node dissection, and coverage in a single surgical session.

1.3. In the operating room

Resection of the tumor in a single block with 1 cm margins. Upon exploration, an infiltrating tumor of the underlying soft tissue (muscular and fatty) was found, without involvement of the ribs, extending toward the axillary hollow without pedicular involvement.

1.4. Left axillary lymph node dissection

The intraoperative examination revealed a morphological appearance of a dermatofibrosarcoma of Darrier and Ferrand with clear surgical margins: 1 cm from the axillary margin, 0.9 cm from the lower margin, 1.3 cm from the medial margin, 1.5 cm from the lateral margin, and 2 mm from the deep margin. Axillary lymph node dissection: 8N-/8N.

It was decided to widen the margins to 5 cm immediately

Coverage was then performed with a latissimus dorsi flap combined with a skin graft. (fig 2) (fig 3)

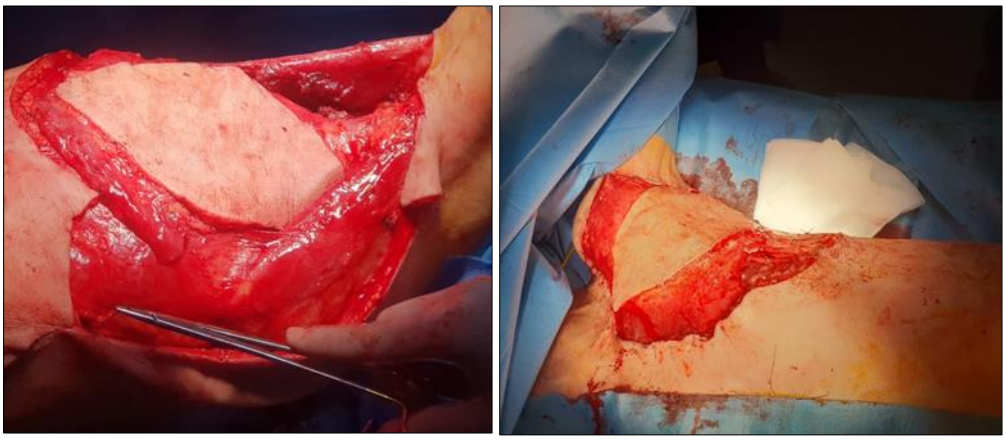

Figure 2 Surgical elevation of the latissimus dorsi flap

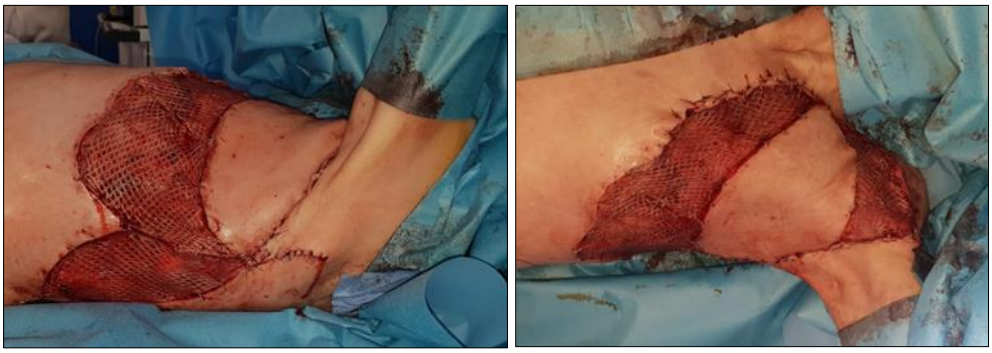

Figure 3 Coverage of the remaining tissue loss with a skin graf

The immediate post-operative course was straightforward with close monitoring of the flap. Dressing changes were done daily with no signs of flap distress or local infection. The graft was mostly successful, and the donor site was monitored with directed healing, achieving complete healing after 15 days. A radiation therapy consultation was requested: it was decided to proceed with 33 sessions of curative radiation therapy over a two-month period. At the end of the sessions, the patient was declared cured, with follow-up appointments every 3 months during the first year.

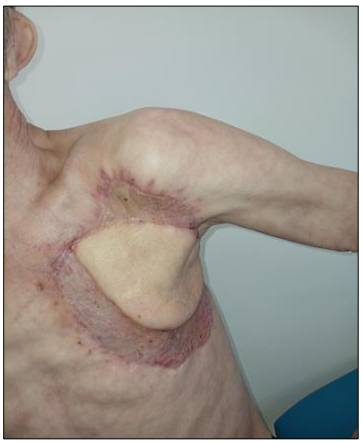

Figure 4 Clinical appearance at 3 months post-surgery

2. Discussion

The differential diagnosis between schwannoma and dermatofibrosarcoma of Darrier and Ferrand can be challenging due to their similar clinical presentation. They typically appear as painless, slow-growing cutaneous or subcutaneous nodules. These tumors can remain asymptomatic for years, making early identification difficult. The integration of histopathology and immunohistochemistry is crucial for establishing an accurate diagnosis. [1]

Despite their clinical similarities, the treatment and prognosis of these tumors differ significantly: Schwannomas require complete surgical excision. Recurrences are rare if the margins are respected. Dermatofibrosarcoma of Darrier and Ferrand, while having a low metastatic potential, is locally aggressive with a high risk of recurrence. Surgery with wide margins is essential. In some cases, adjuvant radiotherapy or targeted therapies like imatinib may be indicated, especially for advanced or recurrent forms. [2] [3]

Thoracic tissue loss presents a significant challenge as its reconstruction aims to restore both structural and functional integrity, while ensuring proper tissue coverage. [4] Directed healing can be an option when the tissue loss is limited and superficial, with a well-vascularized bed. However, this requires appropriate, regular, and thorough dressing changes. The risk of retraction is high. [4] [5]

Direct suturing remains simple and feasible for very small tissue losses. A semi-thick skin graft can be considered for extensive skin loss, but only if there is good-quality granulation tissue. It is essential to secure the graft properly and immobilize the patient post-operatively. [4] [5]

Random skin flaps can be an alternative, but only if the length-to-width ratio is 1:2, and the fascia is included to ensure the flap's viability. [4] [5]

The deltopectoral flap with an internal pedicle is a very simple and reliable option, useful in specific situations when other options are unavailable or exhausted. It is a pedicled flap based on the direct cutaneous perforators of the first three intercostal spaces, supplied by the internal mammary artery. The donor area is grafted at the end of the procedure. [6]

The pectoralis major flap is useful for tissue loss in the median thoracic region. It is a musculocutaneous flap, type 5 in the Mathes and Nahai classification, with the dominant pedicle being the acromiothoracic artery, a branch of the axillary artery. Its pivot point is located beneath the middle of the clavicle, allowing for a 180° rotation. However, this flap may result in moderate functional sequelae. [7]

The latissimus dorsi flap is one of the most commonly used flaps for coverage in this region, as in our case. It is characterized by its ability to cover large areas of skin loss and address certain functional deficits. It is a type 5 musculocutaneous flap in the Mathes and Nahai classification, with the dominant pedicle being the thoracodorsal artery, a branch of the subscapular artery. The pivot point of the dominant pedicle is located at the apex of the armpit. A skin paddle can be raised from the dominant pedicle, with a maximum width of 25 cm and a length of 35 cm. Care should be taken not to extend more than 5 cm below the upper border of the iliac crest. The donor area of the flap is self-closing if it does not exceed 10x12 cm. This flap is very reliable and simple, performed in a single surgical stage, allowing coverage of large thoracic skin losses. Additionally, it tolerates postoperative irradiation well, starting from the second day. [8]

The greater omentum can be indicated for septic lesions or to fill dead spaces.

Parietal reconstruction can be performed using synthetic materials such as Prolene or Gore-Tex, which are easy to apply but carry a risk of infection due to their foreign body nature, or biological materials like the fibula or iliac crest, which make the procedure more complex and lengthier but are more resistant to infection. It is not indicated when the defect is <5 cm with one or two adjacent ribs, or <7 cm at the apex or subscapular region. Additionally, it is recommended after extensive parietal resections or when the scapula is exposed, as it may become incarcerated in the thorax during arm movement. This approach was not used for our patient. [5]

Skin expansion can also be used on the thorax through the insertion and gradual inflation of a silicone balloon with saline solution. This technique allows for minimal scarring while preserving the ideal texture and color of the skin. However, it involves multiple stages, which is a significant disadvantage in terms of coverage after tumor excision.

Lipofilling, or autografting of adipose tissue, remains a valuable option in this context to fill any potential depression and also to improve the quality of the skin.

3. Conclusion

Schwannomas and dermatofibrosarcoma of Darrier and Ferrand represent two distinct mesenchymal entities with distinct characteristics, but they share common aspects. An accurate diagnosis is essential to ensure appropriate treatment, minimizing the risk of recurrence and improving overall prognosis. A multidisciplinary approach remains key to optimal management of these conditions.

Thoracic tissue loss reconstruction relies on a multidisciplinary approach. Among the methods used are skin grafts and muscle flaps, particularly the latissimus dorsi flap, which is best suited for the region and allows for the restoration of both function and skin condition in the affected area. Management depends on the location, extent of tissue loss, and the patient's general condition. Careful planning and close collaboration between plastic surgeons and thoracic surgeons are crucial to optimize functional and aesthetic outcomes.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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