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CASE REPORT

Solitary Fibrous Tumor of the Pleura with Doege-Potter Syndrome

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ABSTRACT
Solitary fibrous tumor of pleura (SFTP) is a rare tumor. Complete tumor resection is the curative treatment and the key to preventing recurrence. We present here the case of a 65-year-old female diagnosed with a Doege-Potter syndrome who underwent resection of a pleural solitary fibrous tumor.

KEYWORDS: Solitary fibrous tumor of pleura (SFTP), Doege Potter syndrome, Surgery.

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INTRODUCTION

Solitary fibrous tumor of the pleura (SFTP) is a rare neoplasm with mesenchymal cell origins. Hypoglycemia associated SFTP is referred to as the Doege-Potter syndrome (DPS) and is caused by inappropriate secretion of an insulin-like growth factor II (IGF-II), **a pattern seen in fewer than 5% of cases** (1, 2, 3).Here, we described a case of malignant SFTP with DPS.

CASE PRESENTATION

A 65-year-old woman with no particular medical history, was hospitalized with a complaint of syncopal episode associated with right chest pain. Clinical examination revealed diminished breath sounds in the right lung field and dullness to percussion.

A chest X-ray showed a right dense, homogeneous, opacity (Figure 1). Blood tests revealed severely hypoglycemia. Chest computed tomography (CT) scan demonstrated a right large, well-circumscribed, homogeneous pleural mass measuring 17 * 13 * 12 cm (Figure 2).

To determine the origin of this tumor, a transthoracic puncture was performed, revealing a pleural origin of the neoplasm.

Right posterolateral thoracotomy through the seventh intercostal space was performed for the resection of the tumor. Surgical exploration revealed a large, well-circumscribed encapsulated pleural mass measured 20 cm and weighed 2000 g (Figure 3).

A complete resection of the mass was performed with a good postoperative evolution including the disappearance of hypoglycemia.The patient was discharged on postoperative day 5.

The histopathologic analysis confirmed the presence of malignant SFTP with free marginal resection. Three years postoperatively, there is no clinical or radiologic evidence of tumorrecurrence.

DISCUSSION

SFTP is a rare primary neoplasm, representing less than 5% of all pleural tumors (1, 2). These **tumors** occur all age, with a peak age of 60-70 years and an even distribution between genders (1, 4).The etiological factors are unknown and no risk factor has been identified yet (1). More than 80% of all SFTP are benign (2, 6).

The majority of these SFTP are usually asymptomatic and are incidental findings. However, 50% of the patients may present with dyspnea, cough, and pleuritic chest pain. These tumors are **associated** with paraneoplastic events, such as hypoglycemia or hypertrophic pulmonary osteoarthropathy **in 5% and 35% of patients respectively** (1, 4, 5).

Hypoglycemia, called Doege-Potter syndrome, has been reported in up to 5% of cases and has been related to hypersecretion of insulin like growth factor **(5)**.

CT scan is the imaging **study** of choice that can identify a smooth, well-circumscribed, homogeneous mass with or without necrosis or calcifications (1).

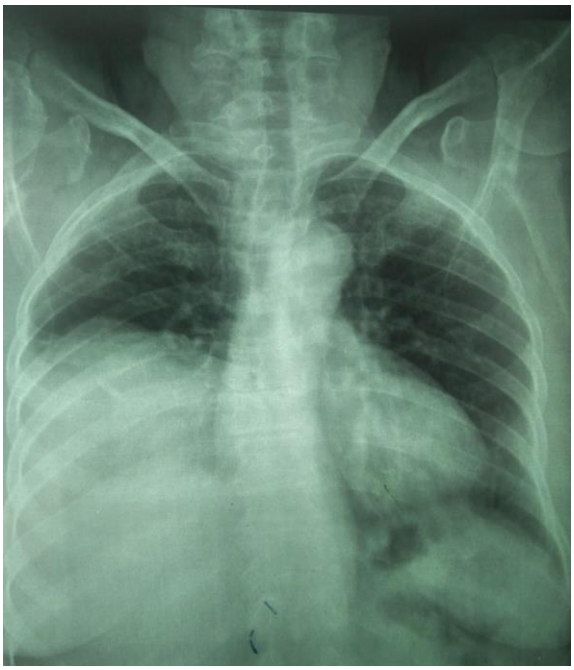

Figure 1: Chest X-ray showing a right lower opacity.

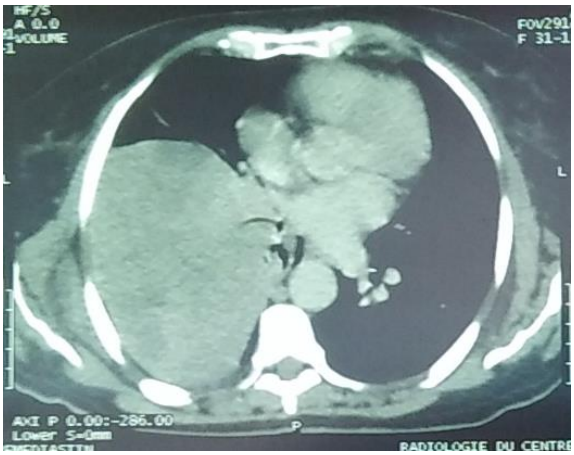

Figure 2: Horizontal chest CT scan in mediastinal section demonstrating a right large pleural mass.

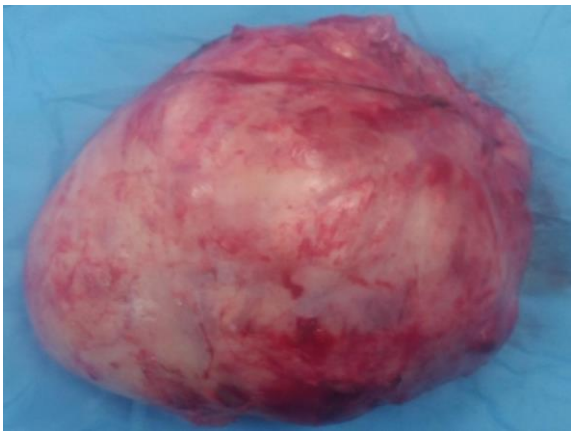

Figure 3: A larger encapsulated SFTP.

Therefore, a complete surgical resection with a margin of 1 to 2 cm of healthy tissue remains the gold standard treatment and the key to preventing recurrence of SFTP (2, 3, 6). Paraneoplastic syndrome usually disappears after surgery (3).

Histopathologically, it arises from the submesothelialmesenchymal layer of the pleura. Immunohistochemically, SFTP is positive for vimentin, bcl-2, CD 34, CD99, but negative for keratin, EMA, and S-100 protein (3,6).

The criteria of malignancy reported by England et al includehypercellularity, pleomorphism with cytonuclearatypia, high mitotic rate, focal necrosis, associated pleural effusion, atypical location, and invasion of adjacent structures (2, 3).

The important prognostic factors are size, abundant cellularity and complete resection with microscopically free margins (3, 4).

Despite that, a routine long-terme follow-up examinations **is** strongly recommended for early detection of recurrence (2,3).

CONCLUSION

Radical **surgical resection** remains the treatment of choice for SFTP and the key to preventing recurrence.

AUTHORS' CONTRIBUTIONS

The participation of each author corresponds to the criteria of authorship and contributorship emphasized in the Recommendations for the Conduct, Reporting, Editing, and Publication of Scholarly work in Medical Journals of the International Committee of Medical Journal Editors. Indeed, all the authors have actively participated in the redaction, the revision of the manuscript, and provided approval for this final revised version.

PATIENT'S CONSENT

Written informed consent was obtained from the patient for the publication of this case report.

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