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Pierre-Marie-Bamberger syndrome – A paraneoplastic syndrome of solitary fibrous tumour of the pleura

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CASE REPORT

The authors report a case of a 75-year-old woman who presented to the Emergency Department with dyspnoea, productive cough, vomiting and unquantified weight loss. She denied fever, night sweats or other symptoms. She had a past history of dementia, dyslipidaemia and right total hip replacement. No relevant family history was known. On physical examination, the patient was fully awake, relatively mobile and disoriented to place and time. Her vital signs were within normal limits with an oxygen saturation of 94% on room air. Lung examination revealed absence of breath sounds over the right hemithorax with increased dullness to percussion. Bilateral pitting oedema extending to the joints was seen in the lower extremities. She complained of pain while palpating both femurs. Digital clubbing was also notable (Figure 1A). Laboratory findings were as follows: haemoglobin 13.7 g/dL (normal, 12–15), mean corpuscular volume 82.6 fL (normal, 83–101), leucocytes 13.7x10^9/L (normal, 4–10), platelets 310x10^9/L (normal, 150–400), lactate dehydrogenase 442 U/L (normal, 125–220), C-reactive protein 4.97 (normal, 0–0.5); with no other relevant findings. Chest radiograph report described a significant right diaphragmatic elevation (Figure 1B). Thoracic and abdominal computed tomography (CT) showed a 17.4x14x19.5 cm heterogeneous mass occupying almost the whole right hemithorax, with areas of necrosis, causing right main and lobar bronchial deviation; small right pleural effusion and liver cysts (Figure 1C). Frontal pelvis radiograph showed multilayered periostitis involving both femoral metaphysis and diaphysis (Figure 2). Bronchoscopy was performed and revealed strictures involving the right main and superior lobar bronchi, hindering the progression of the bronchoscope. Transbronchial biopsies were done through the edematous right superior lobar bronchus and revealed basal cell hyperplasia, without neoplastic cells.

Percutaneous CT-guided biopsy was then performed. Histopathologic examination revealed a fibrous tumour,

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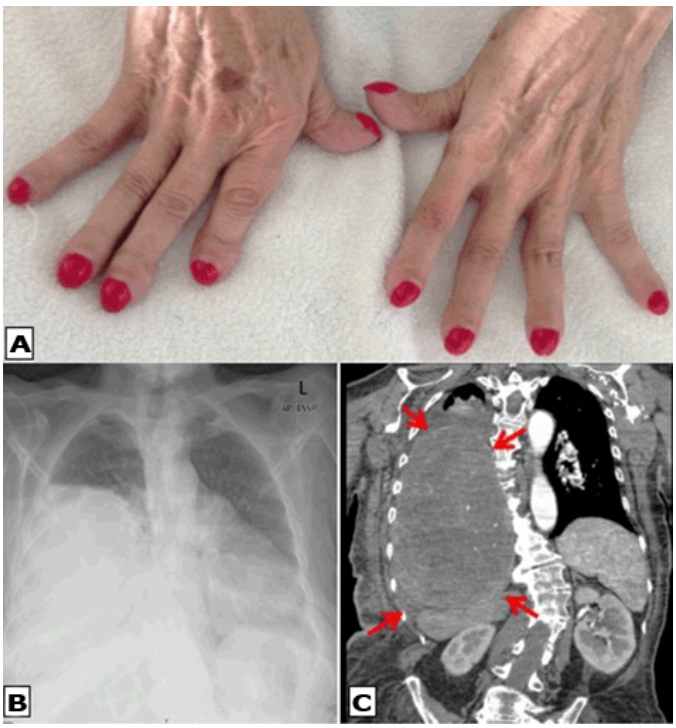

Figure 1: (A) Digital clubbing, (B) Anteroposterior chest radiograph image demonstrates a right hemithorax opacification with contralateral mediastinal shift, (C) Coronal volume-rendered CT image showing a 17.4x14x19.5 cm heterogeneous mass in the right hemithorax, with areas of necrosis.

with negative immunohistochemistry for keratin, S-100 protein, alpha-smooth muscle actin and positivity for CD34, vimentin, bcl-2 and for Ki-67 in 10% of cells in hypercellular areas.

The hospital stay was complicated with recurrent hip prosthesis dislocation, which led to severe functional restriction. As physical rehabilitation was nearly impossible, multidisciplinary treatment decision was to provide palliative care.The patient died of hospital acquired pneumonia seventy-two days after diagnosis.

DISCUSSION

Solitary fibrous tumours (SFT) are relatively rare mesenchymal tumours of fibroblastic origin that can affect any organ [1]. SFT of the pleura (SFTP) account for less than 5% of all pleural tumours [2]. They affect adults in their sixth and seventh decades of life and typically present as incidental asymptomatic slow-growing masses [1]. Larger tumours are more likely to present with symptoms, like cough, dyspnoea, pleuritic chest pain and haemoptysis due to compression of a bronchus [1]. SFTP may be associated with secondary hypertrophic osteoarthropathy also known as Pierre-Marie-Bamberger syndrome, characterized by digital clubbing, periostitis and arthritis, which is thought to be related to increased production of hyaluronic acid by tumour cells [1, 3]. Although it is most commonly associated with non-small cell lung cancer, specifically adenocarcinoma (reported in 0.7–17%), a percentage of up to 20% of SFTP present with this paraneoplastic syndrome [4]. Most of SFTP are benign, although some behave aggressively [1]. Surgical resection via thoracotomy is the mainstay of treatment for most localized SFTP [1]. Resection of giant-sized tumours, such as the one described herein, can be more challenging, due to poor exposure and visualisation, as well as significant blood supply to the tumour [1, 2]. Treatment of secondary hypertrophic osteoarthropathy is best achieved by definitive treatment of the primary pathology, whenever possible [3, 4].

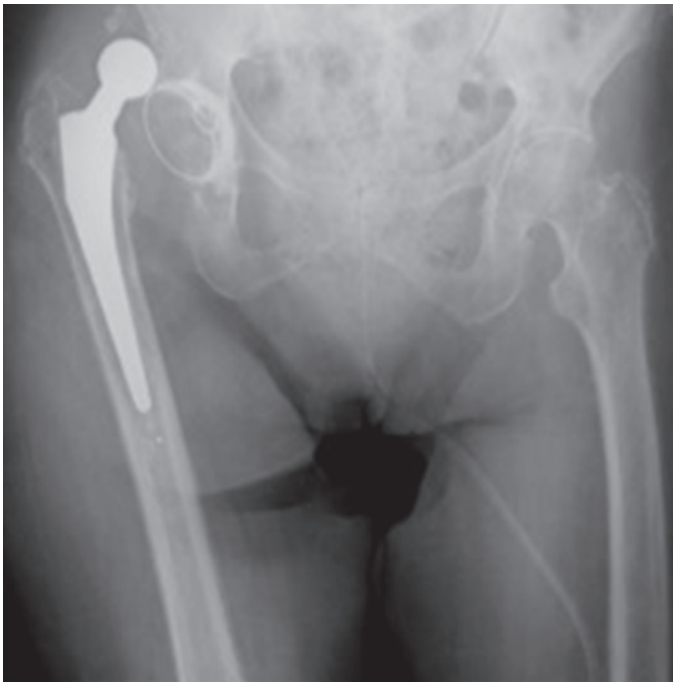

Figure 2: Frontal pelvis radiograph shows right hip prosthesis dislocation and multilayered periostitis involving the femoral metaphysis and diaphysis.

CONCLUSION

Pierre-Marie-Bamberger syndrome is a rare paraneoplastic syndrome, which is rarely associated with SFTP. In a patient with clinical diagnosis of clubbing and joint or bone pain, attention must be directed to chest imaging, even if asymptomatic. Definitive clinical management of this paraneoplastic syndrome is aimed at treating the underlying cause, whenever possible.

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Author Contributions

Sara Neves Sintra – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, revising it critically for important intellectual content, Final approval of the version to be published
Carlos Filipe – Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Adélia Simão – Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
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Guarantor of Submission

The corresponding author is the guarantor of submission.

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Consent Statement

Written informed consent was obtained from the patient for publication of this clinical image.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.