Paraneoplastic Raynaud's phenomenon revealing lung adenocarcinoma

Phénomène de Raynaud paranéoplasique révélant un adénocarcinome pulmonaire

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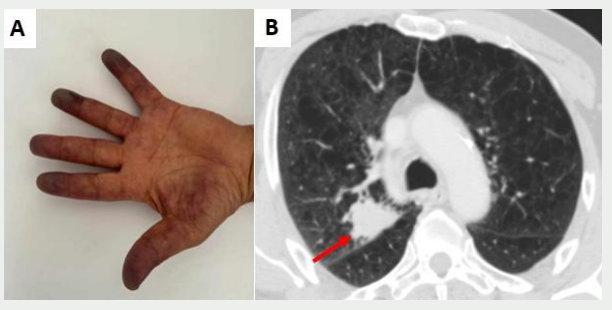

A 69-year-old man, a former smoker with a 40 pack-year history who quit 20 years prior, presented with cyanosis, pain, and paraesthesia in the fingers of his left hand. His medical history included diabetes mellitus, arterial hypertension, dyslipidaemia, and medically treated stage E acute pancreatitis seven years earlier. On examination, the patient was in good general condition, eupnoeic, and had normal oxygen saturation on ambient air. Cardiopulmonary auscultation was normal, and blood pressure was symmetrical in both upper limbs (130/75 mmHg). Examination of the left hand revealed marked coldness and cyanosis in all five fingers' distal phalanges (A), with hypoesthesia in the pulp of the second and third fingers and the thenar eminence, but no motor deficits. Radial and humeral pulses were bilaterally present. No other signs of connective tissue disease or vasculitis were noted. Laboratory findings showed an elevated C-reactive protein level (27 mg/L; normal range: 0–5) with normal complete blood count, coagulation, renal, and hepatic function tests. The clinical presentation raised suspicion of Raynaud's phenomenon, prompting further investigation. Capillaroscopy revealed dilated loops with grade 2 dystrophy. Doppler ultrasound excluded thoracic outlet syndrome and proximal or distal arterial stenosis in the left upper limb. Antinuclear antibodies and antineutrophil cytoplasmic antibodies immunological tests were negative. Based on these findings, paraneoplastic aetiology was considered likely. A thoraco-abdomino-pelvic and cerebral computed tomography scan (B) identified a 33 × 22 mm mass in the apico-dorsal segment of the right upper lobe, contiguous with a bilobed nodule measuring 13 × 7 mm (red arrow). The mass was abutting the major fissure without evidence of invasion. Mediastinal adenopathies were noted, including a 20 × 10 mm subcarinal lymph node and pre-carinal and right latero-tracheal chain nodes. Bronchoscopy findings were normal. Cardiovascular surgeons initiated curative-dose anticoagulant therapy with aspirin and sulodexide, resulting in improvement in trophic disorders. Following a multidisciplinary thoracic oncology meeting, a right pneumonectomy with lymphadenectomy was performed. Histopathological examination confirmed a predominantly solid invasive adenocarcinoma measuring 6 cm, encroaching on the scissura, infiltrating the visceral pleura, and containing endovascular tumour emboli. Postoperatively, the patient experienced worsening of his acrosyndrome, with increased cyanosis and pain in the affected fingers. Doppler ultrasound revealed arterial stenosis, necessitating surgical revascularisation. Despite intervention, his condition deteriorated due to nosocomial pneumonia, requiring intubation and

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mechanical ventilation. The patient passed away shortly thereafter.

Raynaud's phenomenon is an idiopathic ischaemic condition primarily affecting the hands and feet, seen in about 5% of adults, particularly young women [1].It may be associated with connective tissue diseases like scleroderma and systemic lupus erythematosus, as well as conditions such as embolisms, haemostatic abnormalities, trauma, and the use of vasoconstrictive medications [1]. In our case of atypical Raynaud's phenomenon, we excluded various potential causes: autoimmune origins were ruled out by negative immunological tests, drug-induced factors were unlikely due to the absence of vasomotor-active medications, cardiac causes were excluded by a normal cardiac ultrasound, and vascularitis was dismissed based on negative immunological results and capillaroscopic findings. Ultimately, a paraneoplastic origin was confirmed through exclusion and imaging findings.

Paraneoplastic Raynaud's phenomenon is a rare variant associated with malignancies, distinct from the classic idiopathic form [2]. It typically occurs around the age of 53 and affects both sexes, with a slightly higher incidence in men (30%) [3]. It is associated with adenocarcinomas (lung, breast, digestive) in 46.7% of cases [2], as in our case, followed by haematological malignancies [2]. Asymmetrical involvement of the digits is common, with over 80% of cases progressing to ischaemia, necrosis, pulp atrophy, and gangrene [4]. A retrospective analysis of 15 cases found that the median interval between cancer diagnosis and the onset of acral necrosis is two months (range: 1 to 9 months) [2]. In our case, paraneoplastic acrosyndrome was the initial manifestation that prompted the diagnosis of lung cancer. Several mechanisms have been proposed, including tumour invasion of the sympathetic nervous system, microembolism of tumour fragments, increased blood viscosity, and excessive production of vasoconstrictive, thrombogenic, and immunological mediators by tumour cells [5]. Paraneoplastic acral vascular syndrome progresses similarly to other paraneoplastic conditions with most cases showing improvement after chemotherapy, radiotherapy, or surgical intervention for the primary malignancy [5]. Most cases resolve once the underlying tumor is treated [1,5]; however, in our case, the acrosyndrome worsened postoperatively. Prostaglandins, which were not prescribed, might have provided additional relief [2]. Despite its educational value, this report is limited by its single-case nature and lack of generalizability. Furthermore, the acute postoperative deterioration and fatal outcome could not be thoroughly evaluated or addressed due to the accelerated clinical course.

In conclusion, malignancy evaluation is essential for smokers and elderly patients presenting with unexplained or sudden-onset paraneoplastic Raynaud's phenomenon. Early detection and treatment can significantly reduce the risk of digital complications [5].

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