A giant angiokeratoma of Fordyce: an uncommon cause of acute scrotum

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DESCRIPTION

A 34-year-old man presented to the emergency department with intense, sudden and persistent scrotal pain, worsened by the orthostatic position. He denied other accompanying symptoms such as bleeding or history of recent trauma. He referred an asymptomatic congenital angioma of the scrotum and denied any other medical history. Physical examination revealed an extensive red-to-reddish blue papules in the right hemiscrotum, with numerous dark red petechiae (figure 1). It was painful on palpation. Ultrasound revealed a general skin and subcutaneous tissue thickening with multiple dilated vessels, without testicular changes. Systemic examination was unremarkable.

Diagnosis of thrombosed angiokeratomas of the scrotum—Fordyce's angiokeratomas—was made. Since it is an acute scrotum, with uncontrolled pain due to extensive areas of thrombosis, he underwent an urgent surgery with complete excision of the angioma, without complications. Histopathological study confirmed the clinical diagnosis (figure 2). Potential precipitants such as intra-abdominal masses, urinary tract tumours, varicoceles, hernias and angiokeratoma corporis diffusum (Fabry syndrome) were excluded. After 2 years, the patient remained asymptomatic.

Angiokeratoma of the scrotum is often a benign and asymptomatic condition.¹ It is a vascular dermatosis characterised by dilated vessels of the superficial dermis associated with epidermal hyperplasia.² The remaining skin should also be carefully examined for other lesions that may be associated with Fabry syndrome, malignant melanoma, melanocytic naevi or other pathologies.^{1 3}

The curiosity of this case is the extensive lesion in a young patient, associated with an unusual

Figure 2 Haemangioma (proliferation of dilated and tortuous vessels) and it should be noted the presence of fibrin thrombi in the lumen of some blood vessels.

symptomatic presentation of intense pain due to vascular thrombosis, without haemorrhage, requiring an urgent surgical intervention.

Learning points

- ▶ Scrotal angiokeratoma of Fordyce often arise in the second or third decade but are most commonly diagnosed in elderly men and is frequently asymptomatic benign lesion.
- ▶ Although the most frequently described first presentation is bleeding, there are a few cases presenting extensive thrombosis with an uncontrolled pain, making it difficult to diagnosis.
- ▶ Should be excluded other possible associated diseases such as angiokeratoma corporis diffusum (Fabry syndrome), intra-abdominal masses, urinary tract tumours, varicoceles, hernias, malignant melanoma and melanocytic naevi.

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Figure 1 Multiple blue-to-red papules and numerous dark red petechiae with a scaly surface located on the right hemiscrotum.

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