Abrikossoff’s tumour: report of a rare case in anal and perianal region

Abstract

The authors report a rare case of a 47-years old man suffered from Crohn’s disease and depression with multiple prominent lesions in the anal and perianal region. The biopsy of these lesions showed the presence of Abrikossoff’s tumour. This tumour is very rare in the anal region and usually the lesions are small (0.5-3 cm) and solitary. The authors report this case because they considered it an interesting case for the localization and the appearance of the lesions.

Key words: Abrikossoff’s tumor, granular cell tumor.

Case report

A 47-years-old man, suffered from Crohn’s disease and depression, was admitted in our General Surgery Division in November 2001. He showed poor clinical conditions, with a right colostomy well-working and an inflammatory area around this colostomy. Multiple prominent lesions were present in the anal and perianal region. These showed irregular surface, reddish colour, hard-fibrous consistence and were confluent and covered with serum-fibrinous exudate (Photo 1).
The Authors, looking at these lesions, supposed a Crohn’s disease complication or a malignant tumor. Clinical and instrumental examination (blood test, Thorax x-ray, Rectoscopy) were negative so it was necessary a biopsy. The histologic diagnosis was Abrikossof’s tumour. It wasn’t possible to perform a complete excision of tumor because the lesions were large and the patient was in a bad state. After a month he died because he committed a suicide.

Discussion

Granular cell tumours were originally described in 1926 by Abrikossoff. He described this tumor as a asymptomatic solitary nodule with smooth surface, sometimtes associated with overlying acanthosis and/or pseudoepitheliomatous hyperplasia. It has slow growth, rarely exceed 3 cm. and his behaviour is usually benign. The tumour’s histogenesis was very controversy and the numerous theories suggested a variable origin from smooth or stiated muscular cells, mesenchimal or neural cells. Today the neural origin, suggested in the first by Feyrter in the 1935, is the most propable. In the 1962 after immunohistochemical and ultrastructural studies Fischer and Wechsler demonstrated an origin from the Schwann cells. Microscopically the tumor consists of large polyhedral cells with eosinophilic granular cytoplasm, pulvulent aspect, and with a small centrelly-placed nuclei. The cytoplasmatic granules are PAS-positive, both before and after digestion with diastase. Immunohistochemically, granular cell tumors show strong nuclear and cytoplasmic staining with S-100, vimentin and CD-68, lisosomial marker (6-7). The S-100 protein is characteristic of Schwann cells and it proves the probable neural origin of these tumours (8-9). The cells contain also cytoplasmic vacuoles, packets of parallel microtubules, better known as angulate bodies and are generally surrounded by incomplete basal lamina and may show poorly-formed cell junctions (Photo 2). The histologic differential diagnosis is difficult because others pathological entities may show similar features as rhabdomyoma, leymioima and some granular cell lesions associated with trauma or surgical injury. Sometimes it can be possible to mistake a granular cell tumor for well differentiated squamous cell carcinoma because Abrikossof’s tumor can show overlying acanthosis and pseudoepithelomatosus hyperplasia. The majority of granular cell tumor have a benign behaviour, the malignancy are rare, accounting for 1-2%. (5-10). The malignant lesions are different from benign for nuclear pleomorphism, hyperchromasia with vesicular nuclei and large nucleoli (11), high nuclear-to-cytoplasmatic ratio and high mitotic index. The treatment of choice is complete surgical excision (1-12), because if the histologic aspect is characteristic the clinical feature is often uncertain (13-14).

References


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