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

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Riassunto

IL TUMORE DI ABRIKOSSOFF: UN RARO CASO A LOCALIZZAZIONE ANALE E PERIANALE

Gli Autori riportano il caso di un uomo di 47 anni, affetto da M.di Crohn e sindrome depressiva, che presentava multiple lesioni prominenti in regione anale e perianale.
L'esame istologico di tali lesioni, sottoposte a biopsia, dimostrò la presenza di un tumore di Abrikossoff.
Tale tumore si manifesta solitamente con lesioni piccole (0.5-3 cm.) e solitarie e la localizzazione anale e perianale è alquanto rara.
Parole chiave: Tumori di Abrikossoff.

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.

Introduction

Abrikossoff's tumor is a relatively rare, mostly benign lesion.
Altough this disease may arise at virtually any site in the body it is more frequent in the oral mucous and especially in the tongue (1). In the respiratory tract is localized more frequently in the larynx, followed in incidence by the bronchus and rarely trachea (2-3). In the gastrointestinal tract account for about 5-10% of all these neoplasms and approximately one third of these occurs in the esophagus (4).
It develops between the second and sixth decades of life, more frequently among women and blacks. (1)
Usually it appears as a unique firm lesion, a sessile or peduncolated nodule, about 0.5 to 3 cm in diameter. Only in the 10-15% of patient may have lesions at multiple sites which can appear synchronously or metachronously.
Malignant tumours are exceeding rare, accounting for only 1-2% of all granular cell tumors (5). They generally have a similar histologic appearance to their benign counterparts and are diagnosed after the appearance of distant metastases.
The Authors describe an 47-years old man suffered from Abrikossoff's tumour; it is considered an interesting case for the rarity of his location, anal and perianal, and for the appearance of the lesions, which can be mistaken for sqamous carcinoma.

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).

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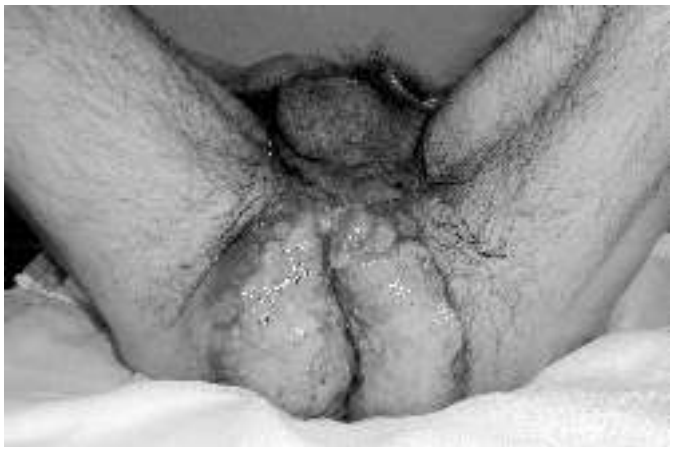

Photo 1: The lesions show irregular surface, reddish colour, hard-fibrous consistence and were confluent and covered with serum-fibrinous exudates.

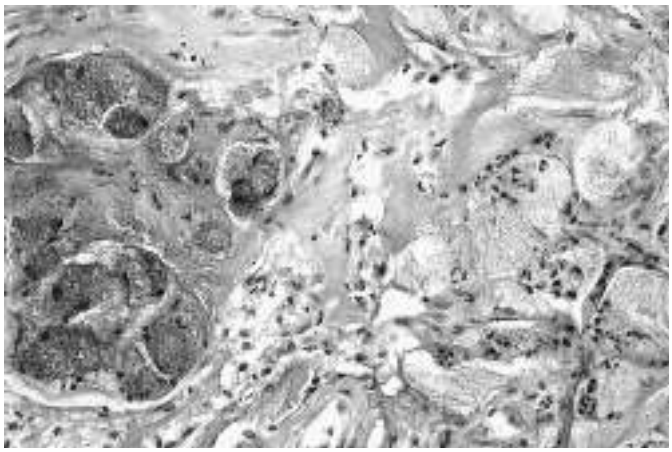

Photo 2: HE (Hematoxilina-Eosina) Tecnique: isthologic image shows a cells proliferations that contain abundant granular cytoplasm, vacuoles, and small nuclei.

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 Abrikossoff'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 suicidie.

Discussion

Granular cell tumours were originally described in 1926 by Abrikossoff.
He described this tumor as a asymptomatic solitary nodule with smooth surface, somentimes 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 muscolar 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 demonstred an origin from the Schwann cells.
Microscopically the tumor consists of large polyhedral cells with eosinophilic granular cytoplasm, pulverulent aspect, and with a small centrally-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, leyomioma 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 Abrikossoff's tumor can show overlying acanthosis and pseudoepitheliomatous 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 vescicular nuclei and large nucleoli (11), high nuclear-to-cytoplasmic 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).

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