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Multiple synchronous granular cell tumours of the esophagus: a case report

Granular cell tumours (GCTs), which were first reported in 1926 by Abrikossoff, who described a series of five tumours in the tongue, are relatively uncommon neoplasms (4). They can be found in many locations including the skin, oral cavity, orbit, breast, external genitals, skeletal muscles, respiratory tract, biliary system and central nervous system (4). Approximately 5-11% of all tumours occur in the gastrointestinal tract and about one third of them appear in the esophagus (5). In all locations, they usually appear as a single lesion, multifocal tumours account for about 7-16% of cases (5).

We present a rare case of multiple synchronous esophageal granular cell tumours.

CASE DESCRIPTION

A 44-year-old man was admitted to hospital on February 12, 1999 for evaluation of an unspecified abdominal pain. He had already been reported with a history of right nephrectomy due to hydronephrosis in 1997. Physical examination did not point to any abnormalities and the laboratory findings were unremarkable. However, routine upper gastrointestinal panendoscopy incidentally revealed three nodules in the esophagus at 36, 38 and 40 cm from the incisor teeth. The tumours were 0.8, 1.0 and 1.5 cm in diameter. They were yellowish, firm and located intramurally. The largest lesion was superficially ulcerated, but the others were covered by intact mucosa. A small antral erosion and deformity of the pylorus were also noted. Endoscopic resection of the nodules was considered, but there was no patient's agreement for this procedure and he was lost during the follow-up.

At endoscopy many tissue samples were taken from the nodules and the diagnosis of granular cell tumours was determined.

Microscopically, in the slides stained with hematoxylin and eosin (H+E), the small nests of large polygonal cells were found in the lamina propria of the mucosa beneath the squamous epithelium that showed the evidence of moderate hyperplasia (Fig. 1) The cells had small hyperchromatic nuclei and

abundant granular cytoplasm. No mitotic activity was found. The cytoplasmic granules were positively stained with periodic acid-Schiff reaction (PAS) and were diastase resistant.

The tumour cells revealed positive immunostaining for S-100 protein using LSAB2/HRP method.

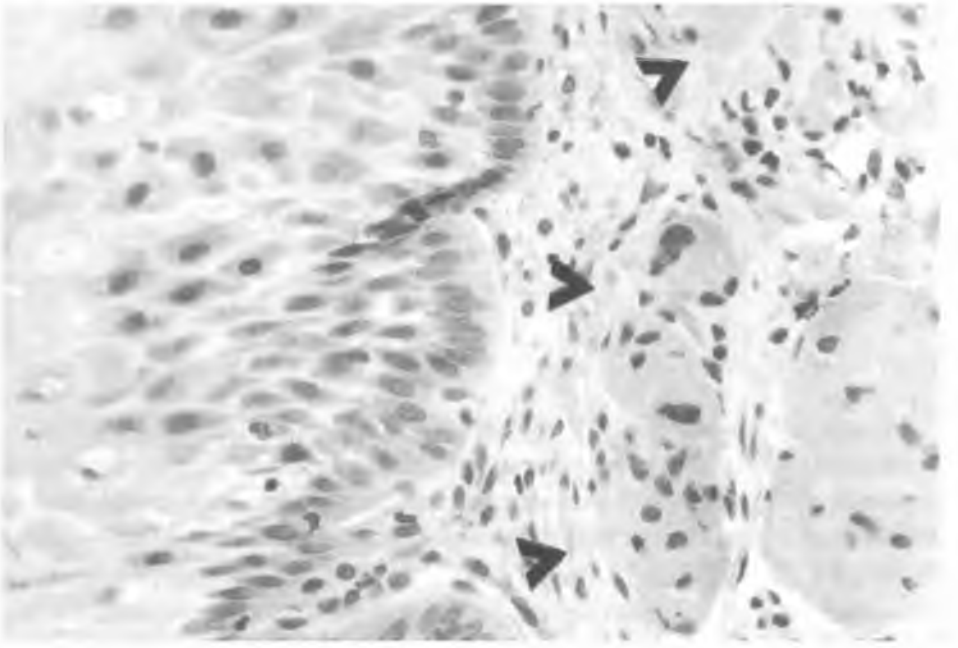

Fig. 1. Granular cell tumour of the esophagus. The nest of polygonal cells with abundant eosinophilic granular cytoplasm and mildly pleomorphic hyperchromatic nuclei in the lamina propria of the mucosa (arrows). Hematoxylin and eosin stain. Magn. x 400

DISCUSSION

Abrikossoff was the first to describe esophageal GCT in 1931 (4). Since then, approximately 200 cases have been reported, with increased frequency due to the widespread use of flexible fiberoptic endoscopy (4). Esophageal tumours are usually solitary, and till now only 30 cases of multiple lesions have been presented in the literature available (1, 2, 3, 4, 5). To our knowledge, the case presented in the study is the 31st to be cited. Most cases are synchronous, but metachronous tumours have been also described (5). It should be emphasised that about one-fourth of the patients with multiple esophageal GCTs had GCTs also in other organs, especially in the skin and stomach (1, 2, 4). Therefore, the examination of more systems is suggested in such patients (2, 4).

Despite the fact of men predominance in esophageal GCTs, there seems to be no sex predilection in the group of patients with multiple lesions (1, 2, 3, 5). The patients' age at diagnosis ranges from 32 to 63 (mean 45) (2, 5). Most multiple GCTs involve the distal and the middle esophagus (1, 2, 4, 5). It is noteworthy, that about 60% of patients with multiple esophageal lesions are symptomatic, suffering from dysphagia, epigastric pain, heartburn or dyspepsia (4, 5).

The diagnosis of esophageal GCTs is mainly based on histopathologic examination of the tissue samples taken at esophagoscopy, but esophageal brushing, typical endoscopic appearance of the lesion and the results of endoscopic ultrasound (EUS) may also be helpful (2, 3, 4). Since the tumours are located intramurally and frequently covered by thick hyperplastic squamous epithelium, many samples should be taken to obtain adequate material for histopathologic examination (4, 5). It is also extremely important to avoid incorrect diagnosis of well-differentiated squamous cell carcinoma, since many GCTs may induce pseudoepiteliomatous hyperplasia of the overlying epithelium (4, 5).

Most esophageal GCTs are benign neoplasms, and only a few cases of malignant tumours have been described (4, 5). They usually remain stable for a long period of time or change slowly, even without therapy (5). Therefore, a conservative therapeutic approach with no removal and close follow--up (endoscopy, histopathological examination) is recommended (4, 5). Surgical or endoscopic excision should be restricted to symptomatic patients, with large tumours (>1 cm) or with histopathological features of malignancy (4, 5). Some authors suggested endoscopic removal of esophageal GCTs after EUS, even in cases with multiple lesions (2, 3).

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SUMMARY

Granular cell tumours (GCTs) are relatively uncommon, usually benign and solitary neoplasms. Approximately 5-11% of all tumours occur in the gastrointestinal tract and about one third of them appear in the esophagus. Till now, only 30 cases of multiple esophageal GCTs have been reported in the literature. We present the case of a 44-year-old man with three synchronous GCTs in the distal esophagus. The lesions were detected incidentally during esophagoscopy. Histopathologic examination of tissue samples revealed the nests of polygonal cells with small hyperchromatic nuclei and abundant granular cytoplasm located in the lamina propria of the mucosa. The cytoplasmic granules were positively stained with PAS and were diastase resistant. The positive immunostaining for S-100 protein was also noted.

Mnogie synchroniczne guzy ziarnistokomórkowe przełyku: opis przypadku

Guzy ziarnistokomórkowe są dość rzadkimi, zazwyczaj łagodnymi nowotworami, występującymi najczęściej jako zmiany pojedyncze. Około 5-11% wszystkich guzów lokalizuje się w przewodzie pokarmowym, z tego 1/3 w przełyku. Do tej pory opisano zaledwie 30 przypadków mnogich guzów ziarnistokomórkowych przełyku. Przedstawiamy przypadek synchronicznego występowania trzech guzów ziarnistokomórkowych w dolnym odcinku przełyku u 44-letniego mężczyzny. Zmiany zostały wykryte przypadkowo podczas ezofagoskopii. Badanie histopatologiczne pobranych wycinków ujawniło w blaszce właściwej błony śluzowej obecność gniazd poligonalnych komórek o małych hyperchromatycznych jądrach oraz obfitej ziarnistej cytoplazmie. Ziarnistości cytoplazmatyczne wykazywały dodatnią reakcję PAS i były oporne na trawienie diastazą. Stwierdzono również dodatni odczyn immunohistochemiczny z przeciwciałem przeciwko białku S-100.