

Granular Myoblastoma: A Case Presentation and Review of Literature

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Summary

The paper describes the unusual localisation of a granular myoblastoma. Granular myoblastoma, or myoblastoma of granular cells, is a tumour of controversial etiology, and according to its rare occurrence in tissues, is an entity in itself. It occurs most frequently in subcutaneous tissues, skin, the breasts, skeletal muscles, and also intraoral tissues, where it occurs most often in the tongue.

In the presented case the tumour occurred on the upper lip, on the border of the lip redness, and after excisional biopsy granular cell myoblastoma was confirmed. According to all criteria the tumour histologically answered the description of the described tumour, and as it was also partially found subcutaneously, pseudoepitheliomatous hyperplasia of the epithelia had occurred. As the alteration was completely excised no further treatment was necessary, and two years after the operation there had been no recurrence.

The reasons for the development of this tumour and its histogenetic origin have still not been completely explained. However new immunocytochemical examinations tend to support the neural theory of its occurrence. Thus, this change should be kept in mind during differential diagnostics of changes which resemble mucocoele, fibromas, pyogenous granulomas, papillomas, angiomas and planocellular carcinomas.

Key words: *myoblastoma of granular cells, upper lip, pseudoepitheliomatous hyperplasia.*

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Introduction

The occurrence of a granular cell myoblastoma, or myoblastoma of granular cells is rare for the physician-clinician and even for the pathohistologist. This tumour of unclear etiology and histogenetic origin occurs most frequently in the

subcutaneous tissues, the tongue, skin, breasts and skeletal muscles (1).

More than 50% of the lesions are situated in the oral cavity (2). Apart from on the tongue, where 35% are found, intraoral cases have been described on the uvala, soft palate, gums and lips (1,2,3,4,5).

The tumour occurs in all ages, most often between 20 - 50 years, equally in both sexes, and more frequently in the black race. It forms a circumscribed painless mass which causes the patient slight discomfort. The tumour has characteristic very slow growth dynamics, and thus in intraoral tissues never grows larger than 0.5 cm in diameter, and in extra-oral tissues never larger than 2 cm (6). Macroscopically they are grey to yellowish-brown nodes. Histologically they are characterised by groups and tracks of large granular cells, 15-20 μ in diameter. The cells contain abundant, acidophilic, pink cytoplasm, surrounding tiny, regular, round nuclei. The cytoplasm contains numerous minute and large granules. The smaller granules resemble lysosomes, although they do not have the characteristic colour for histological staining, while the larger granules are more typical of lysosomes, and apart from other enzymes, also contain acid phosphatase. Although the granules resemble glycogenic granules in the muscle cells, they are not (7). Apart from the granular cells muscle fibres can also be found in the connective tissue stroma, and some granular cells show transition to muscle fibres.

When the changes are found subcutaneously, for some unknown reason, extreme pseudoepitheliomatous hyperplasia occurs, which histologically resembles planocellular carcinoma (1,2,8).

The biological behaviour of this tumour is markedly benign and it is sufficient merely to perform excision up to healthy tissue. Throughout history this tumour has been variously understood; as a focus of cell degeneration, local thesaurismosis, or even a tumour of muscle, neural, fibrous or histiocytic origin (8).

In 1926 Abrikosoff (9) first drew attention to such a lesion. He explained the occurrence of the lesion as degeneration of muscle cells into granular cells as a result of injury or inflammation. The neoplastic theory (2,8) assumes the occurrence of changes from the embryonal muscle cells, and the granules represent embryonal muscle fibril with transverse stripes, according to which the tumour got its name.

The miogenic origin of the lesion (10) has been negated by several reported cases, without skeletal musculature, which in fact negates the possibility of

the occurrence in the skin or gums. The neurogenic theory supports the occurrence of granular cells arising from Schwann's cells (11).

The histocyte theory maintains that the granular cells are in actual fact histiocytes, which contain so far an unknown substance in their granules. (2,5,12).

Case presentation

A male patient, M.J., aged 46 years, was admitted to the Department of Oral Surgery, KBC Zagreb, because of a hard, well circumscribed growth on the upper lip, which was painless but which caused slight discomfort. From case history data it was established that the patient was healthy and that the above change had lasted for approximately two years and was slow growing.

During a clinical examination a yellowish, a hard alteration was found on the edge of the lip, approximately 0.4 cm in diameter, which clinically resembled an atheroma or pyogenic granuloma. A small piece of tissue was obtained by excisional biopsy and sent for pathohistologic analysis, which confirmed granular cell myoblastoma (Fig. 1) with marked pseudoepitheliomatous hyperplasia of the surface epithelia (Fig. 2). The alteration was excised up to healthy (tissue?) and no further treatment was necessary. No recurrence occurred two years after the excision.

Discussion

This type of tumour is rarely seen in dental practice (13), and during a survey of protocol for small operations over a ten year period we failed to find a single case of such a tumour. Thus we can conclude that the occurrence of such a tumour is extremely rare.

From a statistical viewpoint, in our casuistry, excisions of changes on the lips participate with 8%, and of the retroactive/recurring pathohistological findings in the first place are mucocele (pseudocysts of the small lymphatic nodes), followed by fibromas, pyogenous granulomas, papillomas, angiomas, and planocellular carcinomas. These changes should definitely be kept in mind during clinical differential

diagnostics of granular cell myoblastomas (14). It should be stressed that the finding for a histological specimen should be taken sufficiently deeply, and not merely from the surface epithelia, because of the possibility of fatal mistakes in differentiating pseudoepitheliomatous hyperplasia from planocellular carcinoma (15,16,17). Histologically identical tumour cells, such as granular myoblastoma, can be found in congenital epulis (18,19,20). Although histologically epithelial hyperplasia is lacking, it is actually hypoplastic, without muscle cells, and the clinical condition and incidence of tumours is quite different, during differential diagnostics this must also be taken into account. In conclusion it can be said that the origin of this tumour has still not been entirely explained, although many scientists are inclined to accept the neurogenic theory, the search for the cell-precursors of this tumour continues. Immunohistochemical examination of this tumour supports the concept of perineural origin of the tumour because of the presence of carcinoembryonal antigen-CEA (21) which in any case is positive in normal perineural tissue, and is not found in muscle tissue. The neural theory was put forward in 1949 by Fust and Custer (11). They noticed that in numerous cases the granular cells were in close

association with axis cylinders of the peripheral nerves, and that it often appears as though they actually develop from the nerve connective tissue, i.e. Schwann's cells. Today many studies based on immunocytochemistry and electronic microscopy (12,13,21) indicate that the granular cells actually resemble both morphologically and enzymatically Schwann's cells found in cut peripheral nerve. This controversial tumour, for which approximately 19 different descriptive synonyms exist (1), has little clinical significance, apart from during differential diagnostics. Histologically it is clearly recognisable. However, the cause of its occurrence remains unknown. Treatment of this tumour is exclusively surgical, and after adequate treatment there is no recurrence (18).

A review of the literature indicates the rare occurrence of such a tumour (22-25) and all studies are based on presentations of 1-3 cases. Literature gives the number of cases found since 1926 (9) as not more than 1500 cases (26). In our literature Aljinović (26) recorded three cases in one year, although the occurrence of this rare tumour in a period of eight months was considered accidental.