Intracranial trigeminal nerve granular cell myoblastoma

Case report


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The clinicopathological features of an unusual granular cell myoblastoma involving the fifth cranial nerve are presented. Evidence incriminating the Schwann cell as the source of the granular cell is also discussed.

KEY WORDS • trigeminal nerve • granular cell • myoblastoma • schwannoma

Granular cell myoblastoma is a relatively uncommon benign tumor, first described by Abriskossof in 1926. Since then, there have been many conflicting reports regarding its histogenesis.5,9 Recent literature on the electron microscopic findings suggests a schwannian origin of these granular cells,2,5,7,9,11,14 although the striated muscle, connective tissue, and histiocytes have not been totally ruled out.5,11 The commonly used term “myoblastoma” is considered a misnomer by staunch supporters of the schwannian theory of granular cell origin5,7 who regard it as a granular cell neurofibroma or schwannoma.

Granular cell tumors can be found in a wide variety of anatomical locations, the commonest being the tongue,3 subcutaneous tissue, and the mucosa of the upper respiratory tract.11,17 Granular cell tumors involving the central nervous system (CNS) are rare, and such neoplasms of neuropathological interest are classified into four fairly clear subtypes:8 1) asymptomatic granular cell nests, which are seen around the pituitary stalk and are commonly referred to as “choristomas;” 2) supra- and intrasellar granular cell tumors, often presenting symptomatically as space-occupying lesions; 3) intracerebral granular cell tumors of which there are five reported cases (two representing true metastatic lesions and three tumors of unknown histogenesis); and 4) granular cell nests involving peripheral nerves.

We present in detail the clinicopathological features of a granular cell myoblastoma arising from the fifth cranial nerve. This case, which presented clinically as a typical trigeminal neurinoma, is believed to be the first such case in the English literature. A few reports of these tumors arising from the intra-abdominal portion of the vagus nerve6 and other small cutaneous nerves5,11 have been incidental postmortem findings.

Case Report

This 35-year-old woman presented at the neurosurgical outpatient clinic in April, 1979, with complaints of a constant burning sensation in the right temporal region for about 7 years. The sensation was aggravated by talking and mastication, and relieved by analgesic tablets. For the past 2 years, she had noticed a progressive hollowness of the right temporal region associated with difficulty in mastication. She also complained of hypesthesia over the right half of the face. She had developed symptoms of raised intracranial tension in the form of headache, vomiting, and diplopia for the last 15 days. She was not known to be diabetic or hypertensive. No other history was contributory.

Examination. The right corneal reflex was decreased. There was 50% hypesthesia and impaired pain sensation in the area of the first, second, and third divisions of the right fifth cranial nerve. The right temporalis, masseter, and buccinator muscles were atrophic with Grade 2 to 3 power. The jaw deviated to the left. There were no signs of other cranial nerve palsies. The respiratory and cardiovascular systems were normal, and an abdominal examination was unremarkable. The provisional diagnosis was right trigeminal neurinoma. Routine blood and urine examinations were normal. A plain x-ray chest film was normal; however, x-ray examination of the base of the skull
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revealed an eroded and enlarged right foramen ovale (Fig. 1). Right carotid angiography revealed nonfilling of the anterior cerebral artery and a questionable medial displacement of the posterior cerebral artery.

Operation. Through a right temporal craniectomy, subtotal piecemeal removal of the tumor was performed. The tumor was seen arising from the mandibular division of the trigeminal nerve. The mass was firm and encapsulated.

Postoperative Course. The postoperative period was uneventful. The burning sensation and hypesthesia improved; however, the patient developed mild tinnitus and impaired hearing in the right ear. She has enjoyed good health for the past 3 years, with occasional complaints of headache relieved with analgesics. Her only deficit is weakness of the right masseter muscle.

Pathological Examination. The biopsy material was fixed in 10% neutral formalin and processed routinely for paraffin blocks. Sections were cut at 7-μm separations, and stained with hematoxylin and eosin, periodic acid-Schiff (PAS), van Gieson, Masson trichrome, Luxol-fast blue (LFB), and silver stains. A few selected parts were reprocessed from the paraffin blocks according to the method of Vasudev Rao, et al., for cutting semi-thin paraffin sections for better delineation of the morphological characteristics. Other sections were processed for electron microscopic study. Some fragments were deparaffinized, rehydrated, postfixed in 1% osmium tetroxide, and embedded in araldite. These sections were stained with uranyl acetate and lead citrate.

Light microscopy revealed the typical morphological characteristics of a granular cell tumor described earlier by Bangle. The tumor was confirmed to have a fibrous capsule in some of the processed sections. The cells within were seen as irregular strands or groups separated by dense bands of collagen. The contours of the cells were either oval or polyhedral, with a granular eosinophilic cytoplasm. The granules varied in size, but most of them were coarse. The nuclei were relatively small and round, and occasionally included a nucleolus. The chromatin was fine and poorly stained, giving a vesicular appearance to the nucleus (Fig. 2). The granules within the cytoplasm were PAS-positive. Silver impregnation and LFB stains failed to reveal any evidence of nervous tissue.

Electron microscopy revealed poor preservation of cell margins and organelles. However, a few membrane-bound dense granules corresponding to the ones in light microscopy were identified.

Discussion

The term "granular cell" in pathology is merely descriptive terminology to characterize the morphology of cells in any lesion: it does not attempt to identify its histogenesis. Until recently, lesions with a granular cell component were conveniently labeled as "granular cell tumors," regardless of the cell of origin (for example, congenital epulis, xanthomatous lesions, choristomas of the pituitary tissue, myoblastomas, and schwannomas). With advances in electron microscopy and histochemistry, an attempt is being made to clarify the true status of granular cells. However, the basic question of whether they represent true neoplastic cells still remains unanswered.

Granular cell myoblastoma was first described by Abriskossoff in 1926. He considered degenerating myoblasts as the cell of origin, and hence the name "myoblastoma." These tumors commonly arise in organs and tissues derived from mesenchyme or neuroectoderm. The morphological characteristics seen at light microscopy are quite consistent. However, divergent views prevail regarding the histogenesis of these lesions. One school considers them to be an inflammatory, degenerative, or metabolic abnormality, while another suggests that they are true neoplasms. Myoblasts, fibroblasts, histiocytes, undifferentiated mesenchymal cells, and Schwann cells have all been considered as the cell of origin. Recent literature on electron microscopic findings relates granular cell tumors to schwannomas by demonstrating distinct limiting basement membranes with occasional intracytoplasmic axons and myelin figures in both the cell types. Widespaced collagen with a separation of 1200 to 1500 Å is also a common feature in these tumors. The problem of histogenesis is further confused when the tumor is
identified in other than nervous tissue. In such circumstances, it is supposed that they originate from perivascular nerve twigs. Some authors have even reported finding granular cells in normal nerves and in schwannomas. Yet others have demonstrated sequential changes in the conversion of a Schwann cell to a granular cell. Proponents of the schwannian origin theory have recently demonstrated the presence of S-100 protein in granular cells, a protein that is normally present in Schwann cells. This finding further strengthens their claim.

Granular cell tumors of the CNS are rare, and there are few documented cases. Those reported in the cerebral and cerebellar hemispheres are thought to be secondary metastatic lesions, and a case from the spinal leptomeninges is of doubtful cellular origin. Few case reports have described tumors involving either the extracranial portions of the vagus nerve or the smaller subcutaneous peripheral nerves. A thorough search through the English literature failed to reveal any symptomatic granular cell tumor involving a cranial nerve. Hence, ours is the first report of such a tumor. The tumor in our case was symptomatic, with distinct fifth nerve symptoms and signs; at surgery it was identified as arising from the mandibular division of the fifth nerve.

The light microscopic findings in our case were typical of those of similar tumors found elsewhere. The granules were PAS-positive, and groups of cells were separated by dense bands of thick collagen. There were no structures simulating a schwannoma or a nerve. Electron microscopic study of tissue retrieved from the paraffin blocks was attempted, but the cellular details were not satisfactorily preserved due to poor fixation in paraffin processing. However, a few membrane-bound bodies with electron-dense material within them were seen. There were no further details to suggest the histogenesis of the granular cells.

Malignant granular cell myoblastomas are reported in the literature, although only rarely. Our patient had exhibited symptoms for 7 years, but there were no signs of malignancy on histological examination, which proved it to be a benign tumor. The patient had regression of symptoms, with no recurrence 3 years following surgery. We favor a Schwann cell origin for this tumor, but cannot identify whether the Schwann cells are metabolically deranged or if there is a true neoplastic transformation.

Fig. 2. Semi-thin 0.5-μm sections from the biopsy material. Left: Groups of polyhedral and oval granular cells separated by dense bands of collagen can be seen. H & E, × 40. Right: Higher-power view of the granular cells. Note the vesicular nucleus with nucleolus and the coarse granules. H & E, × 100.
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References


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