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Pathology Summit 2018: Granular cell tumor clinical spectrum of the benign and malignant entity - Fariha Kauser, Anshoo Agarwal and Arvind Sinha - Civil Hospital Karachi, Pakistan.

Fariha Kauser, Anshoo Agarwal and Arvind Sinha
Civil Hospital Karachi, Pakistan

Granular cell tumors are rare lesions, although the head and neck region accounts for about 50% of all lesions. The basic cell of origin is now believed to be neural, although previous studies have indicated an origin of the striated muscle or an origin of histiocytic, fibroblasts or pericytes. The tumor usually occurs in middle-aged or older adults. The lesions often show a yellowish pallor or discoloration and usually have a smooth surface. Since most granular cell tumors are benign, surgical excision of the lesion is the treatment of choice. We report four cases of granular cell tumors: Case 1: Imitation in the form of metastatic skin nodule in the umbilical region diagnosed at BP Koirala Institute of Health Sciences, case 2: occurring in a 3-year-old child, case 3: tumor with granular cells coexisting with an epidermoid carcinoma in the respiratory tract and case 4: myoblastoma with granular cells stirring in a burn case, studied at Civil Hospital Karachi. The histopathological results in all these cases showed characteristics of granular cell tumors confirming the diagnosis. Cases are reported for its rarity and unusual presentation.

What is a granular cell tumor?

A granular cell tumor is a rare and generally benign tumor previously known as myoblastoma with granular cells. Their origin has been debated since their first description, but it is now generally believed that they originate from Schwann cells (cells that provide insulation from myelin to the nerves). They are called granular cell tumors because of their appearance under a microscope.

Where does a granular cell tumor occur?

Granular cell tumors occur in the skin or subcutaneous tissue in 30 to 40% of cases. About 50% occur on the head and neck and about 25% on the tongue. They can however occur on any cutaneous or mucous site and sometimes affect the internal organs.

What are the clinical features of a granular cell skin tumor?

A granular cell tumor is most often a solitary, small (usually 1 to 3 cm) and painless tumor. Sometimes mild itching or tenderness is described. They usually appear as a slow-growing skin-colored nodule with a smooth or slightly rough surface.

Who gets granular cell tumor?

Granular cell tumors can occur in anyone, but they tend to be more common in women and people with dark skin. Most tend to develop between the third and fifth decade of life. Multiple granular cell tumors can occur in up to 25% of cases and it is in these patients that associations are occasionally made with other conditions. There appears to be an association with several lentigines, and cases have been reported in patients with an accurate diagnosis of Noonan syndrome, type I neurofibromatosis and Leopard syndrome. Family cases of multiple granular cell tumors also occur.

What tests are needed?

The diagnosis of granular cell tumor is made by skin biopsy. Histopathology reveals characteristic granules in the cytoplasm of large tumor cells. A general medical history and an examination to rule out any potentially associated conditions are helpful.

What is the treatment for granular cell tumor?

Most often, surgical excision of the tumor will provide diagnosis and treatment of choice.

What is the prognosis for a granular cell tumor?

Over 98% of granular cell tumors are benign and must be cured after complete excision. Malignant deviations consume been stated very seldom and tend to be more

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reliably predicted by aggressive clinical features (rapid growth, size greater than 4 cm, necrosis and lymph node involvement) rather than by histological features. Therefore, large or aggressively growing tumors should be followed postoperatively.

A granular cell tumor (GrCTs) is a rare, benign, soft tissue tumor that likely originates from Schwann cells. Although it was originally classified as a myoblastoma, recent studies agree that it is more likely to be of neural origin. GrCTs, although commonly found in the tongue or in the skin and subcutaneous regions, can appear anywhere. GrCTs are rarely found in the extremities. About 0.5% to 2.0% of GrCTs are reported as malignant. A malignant GrCTs is aggressive and has a poor prognosis, while a benign lesion has excellent results after surgical resection. Malignant GrCTs have a high rate of metastases and recurrences, with a mortality rate of 40%. It is therefore important to differentiate a malignant tumor from a benign one. A malignant lesion can be suspected on clinical presentation and confirmed by histopathological examination using the Fan burg-Smith criteria.

METHODS:

This study was a retrospective review of all cases of extremity GrCTs that presented to the Orthopedic Oncology Unit of the University Of Malaysia Medical Center, Kuala Lumpur, Malaysia, from September 2006 to March 2013. The cases were identified from the orthopedic oncology database and the corresponding medical records were found. Histopathology slides were also traced and examined.

A diagnosis of GrCTs can be made easily because these tumors have distinct histological characteristics. However, the origin of cells and its pathogenesis are still under debate. GrCTs were originally called `` granular cell myoblastoma " because the tumors were thought to originate from striated muscle cells. (6) Under microscopy, the tumor cells are visualized as large and of polygonal or round shape, with vesicular nuclei and an abundant eosinophilic granular cytoplasm. The cells are usually separated by thin strips of connective tissue or sclerotic collagen tissue. The overlying skin may experience pseudoepitheliomatous hyperplasia. Currently, the assumption that tumors are neural in origin is widely accepted. Most GrCTs are cytoplasmic and nuclear positive for S-100 and neuron-specific enolase, supporting this hypothesis. GrCTs endure also toughly positive for vimentin and CD68. Tumors are sometimes found near the nerves.

The Fan burg-Smith criteria are a useful tool for diagnosing malignant GrCTs. Its six characteristics of malignancy are presented in Table II. In the present study, one patient was diagnosed with malignant GrCTs. The patient's tumor sample indicated spindle cells, vesicular nuclei with large nucleoli, pleomorphic nuclei and increased mitotic activity (> 2 mitoses per 10 HPF). However, it should be noted that these histological features are subjective and may depend on the pathologist.

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