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OPINION ARTICLE

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PBCC: A Clinical and Histological Overview of a Rare and Invasive Skin Tumor

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Description

Pleomorphic Basal Cell Carcinoma (PBCC) is a rare and aggressive subtype of Basal Cell Carcinoma (BCC), which is the most common form of skin cancer. PBCC is characterized by atypical histological features, including large, pleomorphic, and multinucleated cells. It is a highly invasive and recurrent tumor, and it can metastasize to other parts of the body if left untreated. PBCC typically occurs on the face, neck, and upper extremities, and it tends to affect elderly patients with fair skin. PBCC was first described in 1991 by Weedon and colleagues as a distinct variant of BCC. It accounts for less than 1% of all BCC cases and is considered a rare and aggressive subtype. The clinical presentation of PBCC is similar to other types of BCC, with a slow-growing, painless, and often pigmented lesion. However, PBCC lesions tend to be larger and more aggressive than other types of BCC.

Diagnosis of PBCC

The diagnosis of PBCC is made through a combination of clinical and histological features. The histological features of PBCC include large, pleomorphic, and multinucleated cells, as well as a high mitotic rate and areas of necrosis. Immunohistochemistry can also be used to differentiate PBCC from other types of BCC, as PBCC is typically negative for Ber-EP4 and positive for Ki-67.

Treatment of PBCC

The treatment of PBCC involves complete surgical excision with clear margins. Mohs micrographic surgery is often recommended for PBCC, as it allows for precise and complete removal of the tumor while minimizing damage to surrounding tissue. Radiation therapy may also be used as an adjunctive treatment in cases where complete surgical excision is not possible. The prognosis of PBCC is generally poor, with a higher risk of recurrence and metastasis compared to other types of BCC. The 5-year survival rate for PBCC is estimated to be between 30% and 60%, depending on the stage of the tumor at diagnosis. Factors that are associated with a poorer prognosis include larger tumor size, invasion of underlying structures, per neural invasion, and lymph vascular invasion.

Recent studies have identified potential molecular markers that may be useful in the diagnosis and treatment of PBCC. One study found that PBCC tumors had significantly higher expression of the *p53* protein, which is a tumor suppressor protein that is mutated in many types of cancer. Another study found that PBCC tumors had increased expression of several antigenic factors, including VEGF and bFGF, which promote the growth of new blood vessels and may contribute to the invasiveness of the tumor.

Conclusion

In conclusion, PBCC is a rare and aggressive subtype of BCC that requires careful diagnosis and management. Clinicians should be aware of the clinical and histological features of PBCC, as well as the potential for recurrence and metastasis. Mohs micrographic surgery is the preferred treatment option for PBCC, and recent research has identified potential molecular markers that may be useful in the diagnosis and treatment of this aggressive tumor. With prompt and appropriate treatment, patients with PBCC can achieve good outcomes and long-term survival.

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