Understanding Pleomorphic Dermal Sarcoma: A Guide for Patients

Pleomorphic dermal sarcoma is a very rare skin cancer that occurs in sun exposed areas, especially the scalp. It is thought of as a more aggressive version of atypical fibroxanthoma. It may look like an SCC. Diagnosis is by skin biopsy, though a complete removal of the tumor may be needed for full diagnosis. Treatment includes wide local excision or Mohs surgery. Radiation or systemic therapies are treatment options for more advanced disease or if surgery is not an option. After treatment, monitoring for recurrence and metastasis with regular follow up and imaging is important because these tumors may recur.

Pleomorphic Dermal Sarcoma

Pleomorphic dermal sarcoma (PDS) is a rare but aggressive soft tissue cancer that occurs in sun-exposed areas such as the head and neck. It is thought to be caused by ultraviolet (UV) radiation. Unlike other tumors caused by UV damage like SCC or BCC, PDS is thought to arise from cells that make up the stroma, or supportive tissue, of skin, rather than true skin cells (keratinocytes). It is related to a tumor called atypical fibroxanthoma, but it is deeper and more aggressive. Risk factors include male sex, older age, immunosuppression, lighter skin tone, and history of sun damage or other UV-related skin cancers.

The best way to prevent pleomorphic dermal sarcoma is to protect your skin from the sun. Wearing broad brimmed hats when outside can prevent UV damage to the scalp and head.

Presentation

PDS often occurs on the scalp, forehead or other areas on the head, though it can occur anywhere on the body. It may be mistaken for SCC. It typically looks like a single spot that grows over months and may be skin-colored or slightly red. It might bleed.

Diagnosis relies on a skin biopsy, where a small sample of the spot is removed and examined under a microscope. When a PDS is biopsied, it may be difficult to tell if it is atypical fibroxanthoma or truly a PDS, since these two cancers look very similar microscopically. The diagnosis of pleomorphic dermal sarcoma might be made only after the tumor is *completely* removed, since the diagnosis depends on how deep the tumor invades.

Treatment

Treatment of PDS is usually surgical excision with a wide margin of 1-2 cm. Mohs surgery might also be used if the tumor is in a cosmetically sensitive area. If surgery is not an

option, treatment options include radiation therapy or systemic therapy such as chemotherapy or immunotherapy.

For most patients, the outcome is good after treatment of the tumor. However, between 7-35% of cases may recur at the site of removal. Distant metastasis has been reported in up to 16% of patients. Due to a risk of recurrence or metastasis, close disease surveillance with clinical exams and/or radiological imaging is recommended after treatment.

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