Intracranial extension of an eccrine porocarcinoma

Case report and review of the literature

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Eccrine porocarcinoma is a rare malignant tumor of the true sweat gland. It commonly presents in the lower extremities with lymphatic metastasis. The authors describe the clinical presentation, radiographic evidence, operative discoveries, and pathological findings in a patient with an eccrine porocarcinoma involving the soft tissue of the occiput, which had eroded through the cranium. A review of the literature failed to reveal any other such case. The discussion includes the epidemiology, pathogenesis, treatment, and outcome of eccrine porocarcinomas. The six reported cases of scalp eccrine tumors are reviewed.

KEY WORDS • eccrine porocarcinoma • intracranial lesion

TUMORS of the skin may arise in the surface epithelium or in epithelium of cutaneous appendages. Those arising in the cutaneous appendages may originate from the eccrine gland or have eccrine differentiation. Eccrine porocarcinoma is an uncommon tumor that affects men (55%) and women (45%) almost equally.

Case Report

History. This 78-year-old healthy man presented with a progressively enlarging soft-tissue mass on his occiput. He became aware of the lesion while combing his hair. After 4 years the large size of the mass prevented him from sleeping comfortably. He had no neurological or dermatological complaints. The lesion had not ulcerated, bled, or changed in character. A complete physical, neurological, and dermatological examination was unrevealing. In particular, the skin over the lesion was normal and there was no pre- or postauricular, cervical, or clavicular lymphadenopathy. Prior to referral to this service, resection was attempted by a general surgeon based on the preliminary diagnosis of lipoma; however, the gross tumor appearance and the results of a frozen section changed the differential diagnosis to hemangiopericytoma, melanoma, or plasmacytoma. This unanticipated pathological composition led the surgeons to discontinue the surgery after a biopsy specimen was obtained.

Examination. A computerized tomography (CT) scan was obtained and the patient was transferred to the neurosurgery service. The CT scan demonstrated a soft-tissue mass to the right of midline in the occipital region with lytic bone erosion of the inner and outer table. High-density signal was demonstrated within the tumor and may have represented blood from the initial biopsy. A magnetic resonance (MR) imaging study revealed a dumbbell-shaped lesion that was largely extracranial with a small extension into the cranium. It caused slight deformation of the occipital lobe. On the T₁-weighted MR image the lesion appeared well circumscribed but had mixed signal characteristics (Fig. 1). The hyperintense signal may have been due to blood products from the initial biopsy. The tumor enhanced heterogeneously and there were no cystic structures identified (Fig. 2). Magnetic resonance angiography of the sagittal sinus demonstrated displacement of the sinus inward with flow above and below the lesion but focally absent at the tumor site.

The results of presurgical metastatic and hematological workups, including serum prostate-specific antigen and carcinoembryonic antigen, serum electrophoresis, bone scan, and chest radiography, were negative.

Operation. Nine days after the initial biopsy, the patient was returned to the operating room and the previous incision was opened over the mass. The tumor was well encapsulated, dark brown, and relatively avascular. It was soft and gritty in consistency. The dimensions of the resected mass were 5 × 5.3 cm. Consistent with the MR image, the large extracranial portion had eroded the bone in a circular fashion. The involved bone edges were resected back to normal-appearing cranium. The intracranial component was found to be extradural with a well-defined cleavage plane.

Histopathological Study. Histological evaluation of the specimen revealed a large epithelial neoplasm with connections to the skin surface. The contour was asymmetry.
Intracranial eccrine porocarcinoma

Fig. 1. Sagittal T1-weighted MR image demonstrating the heterogeneous dumbbell-shaped tumor eroding the cranium and mildly deforming the occipital lobe.

Neoplasms of the skin may arise in the surface epithelium or in epithelium of cutaneous appendages. Those arising in the cutaneous appendages may originate from the eccrine gland or have eccrine differentiation. The eccrine gland is a true sweat gland and consists of three parts: the syrinx or straight intradermal duct, the coiled intradermal duct, and the acrosyringium or spiral intraepidermal duct, which opens onto the skin (Fig. 4). A fairly common eccrine tumor is the eccrine poroma, a benign solitary tumor. Eccrine poromas occur most frequently in the palms, soles, axilla, and forehead, where eccrine gland density is highest. The cellular components of an eccrine poroma are similar to those of the normal acrosyringium because this is the structure of origin. Architecturally, the tumors present with a symmetrical endophytic growth pattern that expands the dermis. The associated stroma is often richly vascularized with foci of tortuous and dilated blood vessels. The tumor cells are small and squamous with inconspicuous intercellular bridges, and mitotic activity is absent. Small eccrine ducts or attempts at duct formation are often present. Large focal areas of necrosis may be seen and are not ominous signs. Histological features demonstrated in our tumor specimen that represent malignancy included the irregular cellular shape and size, the irregular cell margins, the tendency to become confluent, and the extension into the subcutaneous tissue.

Fig. 2. Unenhanced (left) and enhanced (right) axial T1-weighted MR images illustrating the heterogeneous nature of the tumor.

Fig. 3. Upper: Photomicrograph at low-power magnification demonstrating the malignant features of this epithelial neoplasm. The aggregations of neoplastic cells vary in size and shape and have irregular contours. They are confluent in areas and extend deeply into the subcutaneous tissue. Lower: Photomicrograph showing hints of tubular formation at high-power magnification (arrowheads).

Pinkus and Mehregan7 described the first malignant tumor developing from the acrosyringium and coined the term epidermotropic eccrine carcinoma. Since then approximately 100 cases have been reported in the literature.
Eccrine porocarcinoma is an uncommon tumor that affects men (55%) and women (45%) almost equally. The mean patient age at presentation is 65.5 years (range 19–94 years). The mean length of time from initial tumor presentation to treatment is 8.5 years (range 2 months–50 years). The tumor can present de novo or arise in association with a preexisting benign poroma, nevus sebaceous, chronic lymphatic leukemia and actinic lesions. It can also arise at radiation ports and sites of trauma. Ironically, the majority of eccrine porocarcinomas develop at sites outside the normal locations of eccrine glands. Instead they are located in the lower extremities (buttocks, legs, and feet) in more than 50% of reported cases. Less frequent sites include the face, ear, and scalp (10%), extremities (approximately 11%), and the trunk and abdomen (approximately 9%). The tumor’s dermatological appearance varies. In some cases, the lesion may ulcerate, bleed, or present as warts. The overall local recurrence rate after excision is 25%. Tumor spread occurs tangentially in the lower one-third of the epidermis and later the tumor infiltrates the dermis, subcutaneous fat, and lymphatic system. Twenty-five percent of patients develop distant metastasis. Common sites include lymph nodes, retroperitoneum, peritoneum, femur, breast, liver, bladder, ovary, adrenal glands, and lung.

The propensity for local recurrence and metastases mandates complete radical excision of the tumor with negative margins as treatment. If regional lymph nodes are involved, a regional lymphadenectomy has been advocated by Goedde, et al. Patients with metastatic and recurrent disease have a poor outcome. Because of the small number of cases, treatment recommendations for these patients are not well established. Both radiotherapy and chemotherapy modalities have been used with mixed response rates. Treatment with retinoids and interferon has been shown to have some benefit in isolated cases.

Review of the literature documents six cases of scalp tumors of sweat gland origin. Hymann and Brownstein described two cases; however, from this report no distinction can be made between benign eccrine poromas and the malignant eccrine porocarcinoma. In two of the remaining four cases the site of the tumor is described as “scalp.” In the final two cases involvement of peristium is described. One patient required a wide skin excision, radical neck dissection, advancement flap, and radiation treatment for two of 38 positive nodes. There is no record of visceral metastases in these case reports. Our case is a unique presentation of an elderly man in whom an eccrine porocarcinoma of the posterior occipit destroyed the occipital calvaria and compressed the occipital lobe. It did not involve the dura or sinuses.

References

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