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Case Report

Proliferating Trichilemmal Tumors: CT and MR Imaging Findings in Two Cases, One with Malignant Transformation

Hyung-Jin Kim, Tae Sook Kim, Kyung-Hee Lee, Young Mo Kim, and Chang Hae Suh

Summary: We report the imaging findings in two patients with proliferating trichilemmal tumors. In the first patient, the tumor arose on the lower lip, a very unusual location for this type of tumor, and showed malignant transformation with metastasis to a regional lymph node. It was seen as a poorly marginated soft-tissue mass with isointense signal on T1-weighted MR images and hyperintense signal on T2-weighted images. Large areas of high signal intensity caused by necrosis were also found within the tumor on T2-weighted images. After IV administration of contrast material, the mass showed significant enhancement, with considerable portions remaining unenhanced. In the second patient, the tumor originated from a preexisting trichilemmal cyst and occurred in the hair-bearing area of the posterior part of the neck. CT scans showed a well-encapsulated cystic mass that contained multiple speckled calcifications in a wall of variable thickness. There were several foci of smooth soft-tissue elevations from the inner wall of the mass, which corresponded histologically to proliferating portions of trichilemmal cyst.

Proliferating trichilemmal tumor (PTT), also referred to as proliferating trichilemmal cyst or pilar tumor, is a benign tumor originating from the outer root sheath of a hair follicle (1). It is usually a solitary lesion and most commonly occurs in elderly women (2). Although considered biologically benign, PTT may be locally aggressive. In rare instances, malignant transformation has been reported, evidenced by regional or distant metastases (3–7).

To our knowledge, imaging findings of this unusual tumor have not been described. We report the CT and MR imaging findings of two cases of PTT, one of which was distinctive because it arose in the lower lip, an unusual location for this tumor, and showed malignant transformation, with metastasis to a regional lymph node.

Case Reports

Case 1

A 75-year-old man presented with a 4-year history of a slowly growing, painful mass in the lower lip. Incisional biopsy performed at an outside clinic 1 year earlier revealed squamous cell carcinoma. After treatment with one cycle of chemotherapy, the patient refused further treatment until the time of his visit to our hospital, which was prompted by recent rapid growth of the mass. On physical examination, a 3.0- × 4.5-cm hard, tender mass was noted in the lower lip. The mucosa of the lip was grossly intact. A 1- × 1-cm lymph node was palpated in the right submandibular area.

With an assumption of lip cancer, MR imaging was performed for staging workup. The MR studies showed a poorly marginated, ovoid, soft-tissue mass in the lower lip. Relative to muscle, the mass showed isointense signal on T1-weighted images (Fig 1A) and hyperintense signal on T2-weighted images (Fig 1B). T2-weighted images also revealed several areas of high signal intensity, suggestive of cystic or necrotic change (Fig 1B). The mass was seen to violate the tissue planes of the lip by penetrating the orbicularis oris muscle. After IV administration of contrast material, the mass showed significant enhancement, with considerable portions remaining unenhanced (Fig 1C). There were several small submandibular lymph nodes in the right side of the neck.

Wide resection of the tumor and bilateral modified neck dissection were performed, and the surgical defect was reconstructed by a radial forearm free flap procedure. Macroscopic sections revealed a 2.7- × 2.0- × 2.0-cm yellowish white, poorly defined, lobulated, solid, firm mass containing an irregular central cavity (Fig 1D). The overlying skin appeared focally granular. Microscopically, the mass was composed of multiple proliferating nests of reserve cells of squamous epithelium showing trichilemmal keratinization (Fig 1E). These cells showed high nuclear-to-cytoplasmic ratios, pleomorphic nuclei, and frequent mitoses (Fig 1F). Large areas of necrosis were found within the tumor, and foreign body granulomas were present in the overlying skin as a result of microrupture of the tumor. The histologic diagnosis was malignant PTT. Of all the lymph nodes resected, only one from the right submandibular chain was positive for malignancy. The metastatic node was composed of atypical squamous cells with the same histologic features as the primary tumor.

Case 2

A 54-year-old woman presented with recent rapid growth of a mass in the neck. The mass had been growing slowly since she had first noticed a small lump 10 years earlier. Physical examination revealed a 10- × 10-cm soft, mobile, fluctuating mass in the left posterior part of the neck. The overlying skin was normal in color and texture. Aspiration cytology performed in the outpatient department showed no evidence of malignancy.

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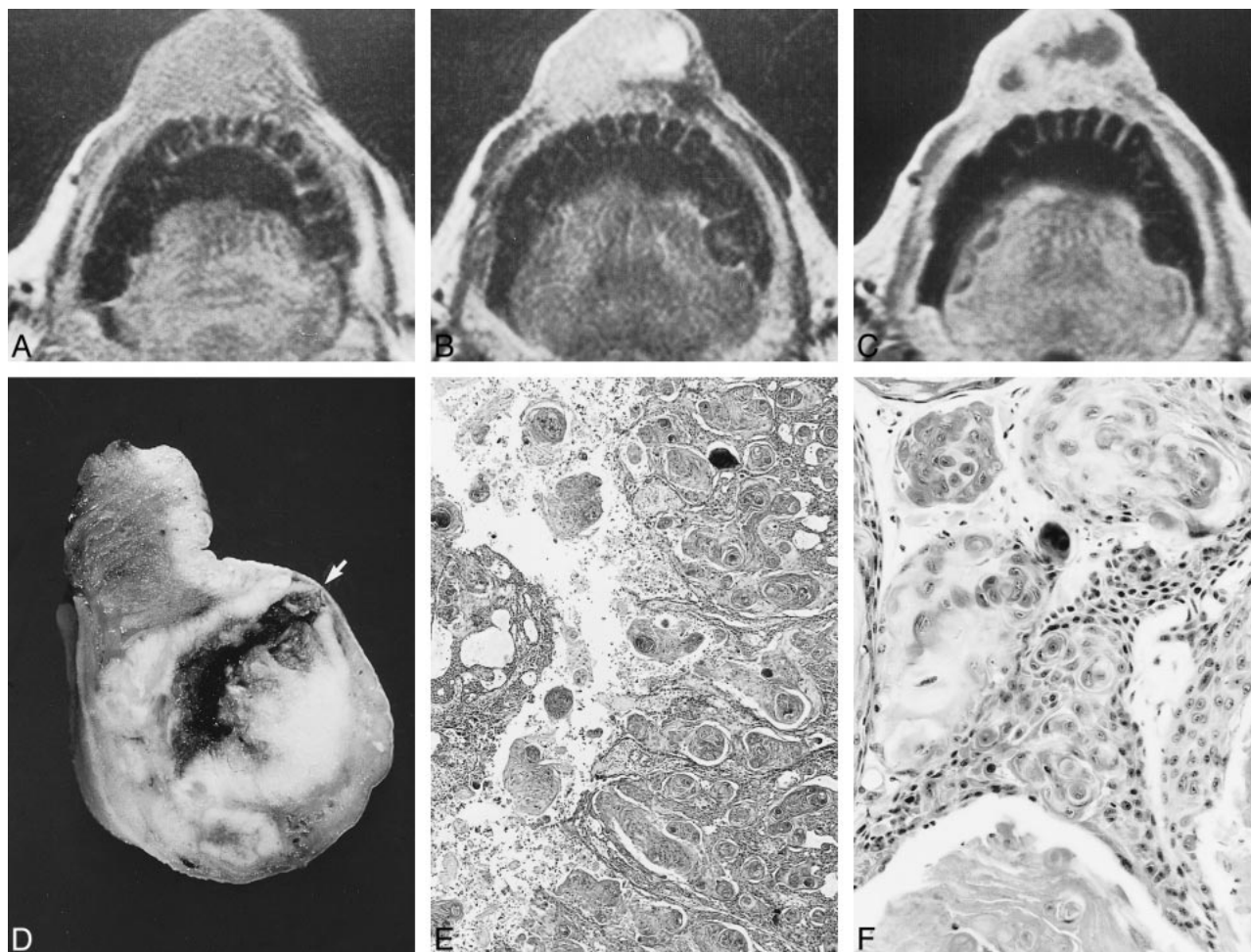


FIG 1. Case 1: 75-year-old man with malignant proliferating trichilemmal tumor.

A, Axial T1-weighted MR image shows a poorly defined soft-tissue mass in the central portion of the lower lip. The signal intensity of the mass is comparable to that of the adjacent muscles.

B, On axial T2-weighted MR image, the mass becomes hyperintense with some portions being much higher in signal intensity. The irregular margin of the mass and its infiltrating nature through the orbicularis oris muscle are better seen on the T2-weighted image.

C, Axial contrast-enhanced T1-weighted MR image shows significant enhancement of the mass, with considerable portions remaining unenhanced (Fig 1C and D). The areas of the tumor showing no enhancement appear larger on the contrast-enhanced image than on the T2-weighted image.

D, Photograph of a cut section shows partially poorly defined, lobulated, soft-tissue mass, which has a large irregular central cavity. The continuity of the mass with the overlying epidermis (arrow) is a general feature of a tumor of hair sheath origin.

E, Photomicrograph shows numerous proliferating lobules of reserve cells with extensive trichilemmal keratinization (hematoxylin-eosin, original magnification $\times 20$).

F, Higher-magnification photomicrograph shows dysplastic cells with high nuclear-to-cytoplasmic ratios, nuclear pleomorphisms, and frequent mitoses. There is no evidence of a granular layer associated with keratinization, which is a useful feature for distinguishing PTT from squamous cell carcinoma (hematoxylin-eosin, original magnification $\times 200$).

Assuming a benign cyst of the skin and its appendage, CT of the neck was performed. Contrast-enhanced CT scans showed a large, well-encapsulated, cystic mass in the subcutaneous fat of the left posterior portion of the neck. The tumor had a well-enhancing wall of variable thickness, which contained multiple, speckled calcifications (Fig 2A), and several foci of smooth soft-tissue elevations arose from the inner wall of the mass. Although the paravertebral muscles were compressed, there was no evidence of extracapsular spread.

Excisional biopsy revealed an 8.0- \times 6.5- \times 7.0-cm grayish white, well-encapsulated, unilocular, cystic mass containing brownish serous fluid, with several smooth excrescences from the inner wall of the mass (Fig 2B). Microscopically, two different histologic features were found in the wall of the mass. Most of the lining of the cyst was composed of epithelial cells, with no clearly visible intercellular bridges. Abrupt keratinization was also seen mainly in the inner layer of the epithelial

lining. This portion of the mass corresponded to trichilemmal cyst. The other component was found in smooth excrescences of the cyst wall. Here, there were multiple proliferating nests of reserve cells, which underwent differentiation into large polygonal keratinocytes with abrupt keratinization (Fig 2C). This portion of the mass corresponded to PTT. Multiple foci of calcification were seen in both components of the mass. The final diagnosis was PTT originating from a trichilemmal cyst.

Discussion

PTT is a neoplasm that probably arises from a preexisting trichilemmal cyst subsequent to trauma or inflammation (8, 9); however, it can also occur de novo, without a preexisting cyst (10). The ambiguity of the origin of this tumor has resulted in

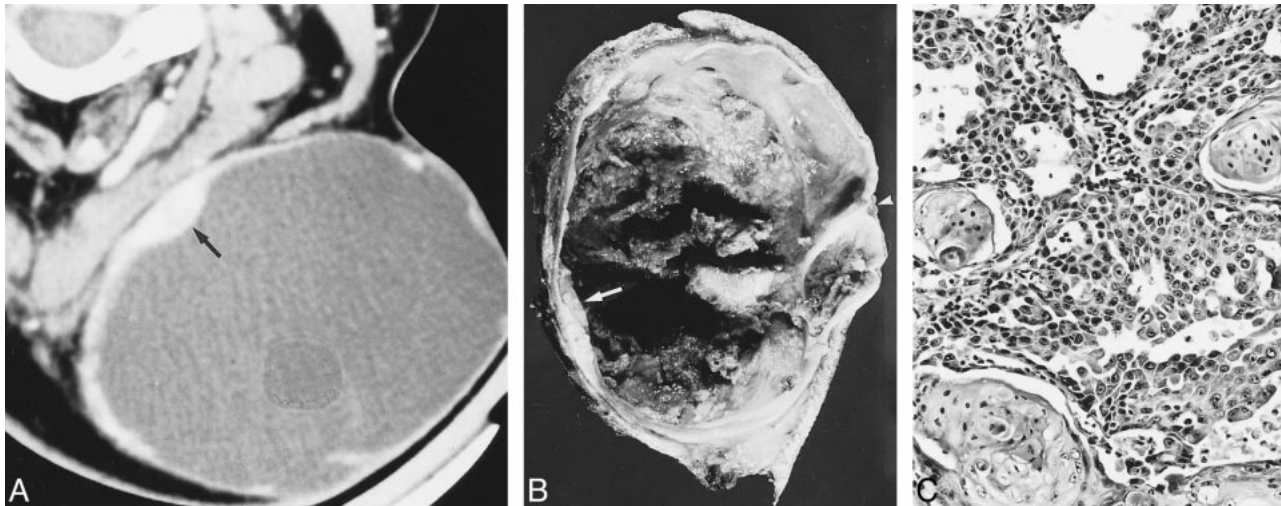


FIG 2. Case 2: 54-year-old woman with coexistent proliferating trichilemmal tumor and trichilemmal cyst.

A, Axial contrast-enhanced CT scan at level of the submandibular glands shows a large, well-encapsulated, cystic mass located in the subcutaneous fat of the left posterior part of the neck. It has a well-enhancing wall of variable thickness, which contains numerous speckled calcifications. Note a focal area of smooth soft-tissue elevation (*arrow*) from the anterior wall of the mass. There were several similar foci of soft-tissue elevations in the inner wall of the mass (not shown). The outer portions of the mass abut the overlying skin. Although the lesion apparently reveals a mass effect on the adjacent paravertebral muscles, the fatty interface is seen to be well preserved.

B, Photograph of a cut section corresponding to A shows a well-encapsulated, unilocular, cystic mass continuous with the overlying epidermis (*arrowhead*). There is a focal, smooth excrescence of soft-tissue (*arrow*) in the deep portion of the inner wall.

C, Photomicrograph of soft-tissue portion of the mass shows extensive areas of trichilemmal keratinization without formation of a granular layer. The reserve cells reveal differentiation toward large polygonal keratinocytes (hematoxylin-eosin, original magnification $\times 200$).

a host of different names to describe it, including carcinoma in sebaceous cyst, proliferating epidermoid cyst, invasive pilomatricoma, trichochlamydocarcinoma, trichochlamydoacanthoma, giant hair matrix tumor, invasive hair matrix tumor of the scalp, trichilemmal pilar tumor, and proliferating trichilemmal tumor (1, 6, 8). Most investigators now consider PTT as a tumor originating from the outer root sheath of a hair follicle (6, 11), and most of the reported cases of squamous or basal cell carcinomas arising in sebaceous cysts are now believed to be PTTs (2, 12).

The usual clinical presentation of PTT is that of a long-standing, subcutaneous, cystic nodule that slowly progresses to a large, nodular mass, often following a history of trauma or inflammation (2). The tumor preferentially arises in areas of dense hair follicle concentrations, and about 90% of cases occur on the scalp, with the residual 10% occurring mainly on the back (1, 3). Other, less common locations include the vulva, nose, mons pubis, buttock, wrist, chest, and elbow (1). Women are affected in more than 80% of cases, and the average age of patients is 65 years (1, 8, 9). The presentation is nearly always that of a single lesion, but, rarely, multiple lesions are seen. An associated trichilemmal cyst with lobules of PTT is also encountered occasionally (1), as in case 2 in the present study.

Grossly, PTT appears as a lobulated, well-circumscribed mass, and the external skin may be atrophied or ulcerated. The cut surface generally has a honeycomb appearance with spaces and small

cysts filled with keratinous material (2). Histologically, PTT is made up of massively proliferating lobules of squamous epithelium showing multiple central areas of trichilemmal keratinization and formation of homogeneous keratin cysts (3, 9). The tumor masses are surrounded by a layer of basaloid cells in which the tumor cell nuclei palisade and the cell membranes rest over an outer layer of PAS-reactive basement membrane. Toward the center of the tumor are larger cells, some having clear or vacuolated cytoplasm containing glycogen granules (9). Areas of necrosis, calcification, and hyalinization may be seen (7, 10). The stroma is usually fibrous and shows a variable inflammatory reaction, including foreign body giant cells (1, 10). The most characteristic histologic feature of PTT is trichilemmal keratinization. Peripheral cells enlarge and become pale-staining and glycogen-rich, with abrupt transition to a dense keratin without the presence of a granular layer (2, 10). Although PTT appears to originate from the outer root sheath of a hair follicle, the occurrence of squamous eddies as well as areas of basaloid cells, hyalinization, and calcification, similar to pilomatricoma, suggest a differentiation not only toward the trichilemmoma but also to the infundibulum and matrix (10). The tumor cells in many areas show nuclear atypia, which at first glance may suggest a false impression of squamous cell carcinoma. Although occasional cells with hyperchromatic nuclei, mitotic figures, and scattered dyskeratotic cells may be present, these do not necessarily indicate malignant transformation (9, 10). Abrupt keratinization, low mi-

totic activity, minimal pleomorphism, and sharp demarcation between the stroma and adjacent dermis are helpful in differentiating PTT from squamous cell carcinoma (3, 10).

Although PTT is generally considered biologically benign, even though histologically indistinguishable from squamous cell carcinoma in some cases, malignant PTT has been reported. Saida et al (13) suggested three stages in the oncologic development of a malignant PTT: the adenomatous stage of the trichilemmal cyst, the epitheliomatous stage of the PTT, and the carcinomatous stage of the malignant PTT. A rare occurrence of PTT with spindle cell carcinoma has also been reported (7). Unfortunately, distinctive histologic or immunohistochemical markers of malignancy do not exist (4). Some recent reports have shown DNA aneuploidy and, in some cases, an increased proliferation index, suggesting that PTT may be a premalignant tumor (14). Rapid enlargement of long-standing nodular scalp lesions and histologic evidence of significant abnormal mitosis, marked cellular pleomorphism, infiltrating margins, and aneuploidy may indicate malignant transformation (1, 9). However, the only unequivocal criterion of malignant PTT is metastasis, either regional or distant, and metastases to cervical and mediastinal lymph nodes, lung, pleura, liver, and bones have been reported (3–7).

To our knowledge, imaging findings of PTT have not been reported previously. This study shows that PTT can manifest as either a cystic or solid mass on imaging studies. When solid, signal intensities of the tumor on MR images were grossly the same as those of most other soft-tissue tumors: areas of hypointensity on T1-weighted images, hyperintensity on T2-weighted images, and substantial enhancement after contrast, as demonstrated in case 1. Poorly defined margins as well as penetration of the tissue planes suggest the malignant nature of the tumor. When cystic, the tumor is typically benign-looking on CT scans, as demonstrated in case 2. Smooth soft-tissue elevations from the inner wall of the mass are an important clue in predicting the nature of the mass, not a pure cyst but a cystic tumor. These areas corresponded histologically to proliferating lobules of epithelium, characteristic of PTT.

The tumor presented in case 1 is worth further mention. It occurred in a man and originated from

the lower lip, which is an unusual location for this lesion. It is not surprising that the patient was initially treated under the diagnosis of squamous cell carcinoma, as a small biopsy specimen may be insufficient to diagnose this kind of sophisticated tumor correctly. Careful histologic examination of the whole specimen is mandatory to differentiate PTT from squamous cell carcinoma. Although objections may be raised as to the true malignant nature of the primary tumor, we believe that metastasis to the submandibular lymph node suffices as evidence of malignant PTT in the present case.

Wide excision of the tumor with a 1-cm conservative margin of normal tissue is the treatment of choice for PTT. Adjuvant chemotherapy is not superior to adequate surgery alone (1). In malignant PTT, the patient should be observed closely to detect any evidence of metastasis.

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