A Case of Chondroid Syringoma with Small Tubular Lumina

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Chondroid syringoma is an uncommon, benign neoplasm of sweat gland origin. It clinically presents as a slowly-growing intradermal or subcutaneous nodule, and is usually observed on the head and neck of middle-aged to elderly people. Histologically, two types of chondroid syringoma can be recognized: one common type has tubular and cystic lumina and the other extremely rare type has small tubular lumina. We herein report a rare case of chondroid syringoma with small tubular lumina. (Ann Dermatol (Seoul) 18(1) 40～43, 2006)

Key Words: Chondroid syringoma, Small tubular lumina

INTRODUCTION

Chondroid syringoma, or mixed tumor of the skin, is a benign neoplasm of the sweat gland which is most often seen on the head and neck[1]. This mixed tumor of the skin was originally defined by Billroth in 1859 as an entity having the same histopathologic properties as mixed tumors of the salivary glands. The term "chondroid syringoma" was first used by Hirsch and Helwig[2] in 1961 because of the presence of sweat gland features in the cartilage-like stroma of this tumor. This benign and rare skin appendageal tumor usually occurs in the head and neck region, and it characteristically presents as a slowly-growing, painless, subcutaneous or intracutaneous nodule, with a 0.5 to 3.0 cm diameter in male adults[1,3]. The lesion is commonly mobile and distinct from the surrounding tissues. The diagnosis of chondroid syringoma is uaually made retrospectively based on histological confirmation, other diagnoses having been considered beforehand. These include sebaceous or dermoid cyst, neurofibroma, dermatofibroma, basal cell carcinoma, squamous cell carcinoma, pilomatricoma, and histiocytoma[4]. Histologically, the tumors are classified into two types: tumors with tubular cystic lumina lined by a double-layered epithelium and tumors with small lumina lined only by a single-layer of epithelium[5]. It is known that the latter is an extremely rare tumor type. Likewise, among the seven cases previously reported in the Korean literature, only one case[6] corresponded to the small tubular lumina type.

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CASE REPORT

A healthy 67-year-old woman presented with an asymptomatic nodule on the right parietal area of the scalp. The lesion had been incidentally noticed two months previously. On admission, a solitary, relatively well-demarcated, 1.5 cm sized, round, flesh-colored, hard nodule was located on the right parietal area of the scalp (Fig. 1). On physical examination, there was no other remarkable finding[5]. Her past history and the family history were not contributory. The histologic examination revealed a well-demarcated tumor composed of epithelial and stromal components. The epithelial component

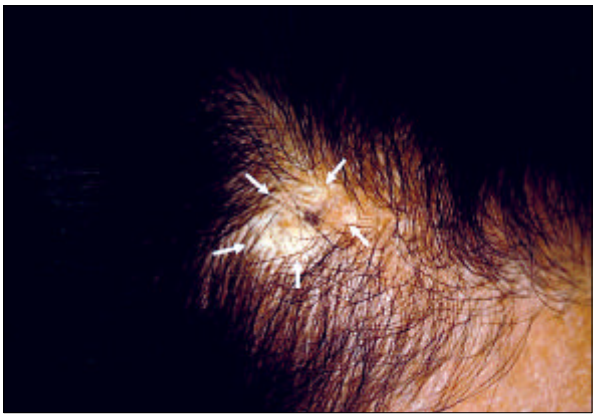

Fig. 1. A relatively well demarcated, skin-colored, firm, 1.5 cm sized, slightly elevated nodule on the scalp.

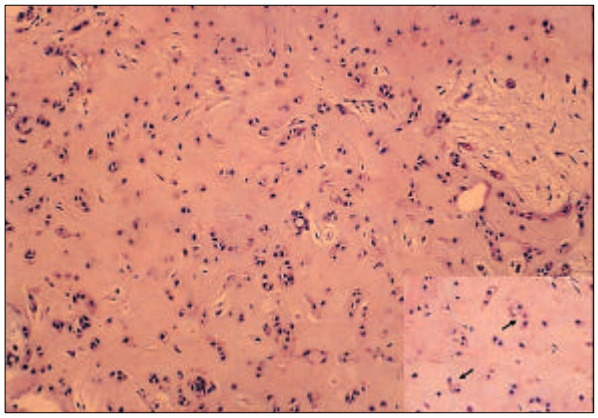

Fig. 2. Biopsy specimen shows basophillic stroma and the tubular alveolar structures which are lined by one layer of cells. Inset: small comma-like proliferations. (H & E, ×200).

showed numerous small ducts lined by a single layer of flat epithelial cells from which small comma-like proliferations often extended into the stroma (Fig. 2). The epithelial cells were positive for S-100 protein upon immunohistochemical examination (Fig. 3) and some of the tubular alveolar structures contained PAS positive, diastase-resistant, eosinophilic material (Fig. 4). The stroma had a mucoid, faintly basophilic substance which was positive for alcian blue stain (Fig. 5). There was no evidence of malignancy, and the lesion was excised completely. The patient remained free of tumor recurrence during the 12 month follow-up period.

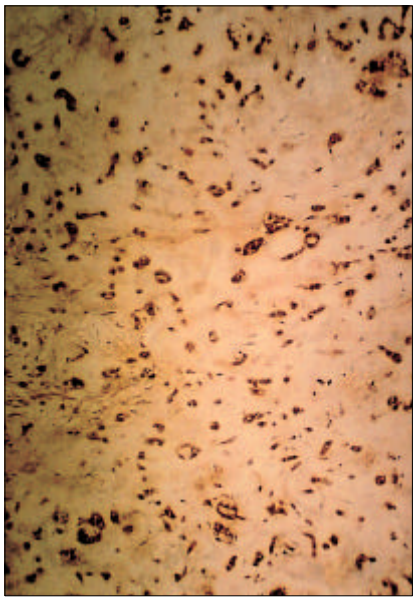

Fig. 3. This figure shows the moderate staining reaction for S-100 protein in the cells of the tubular structures (S-100 protein, ×200).

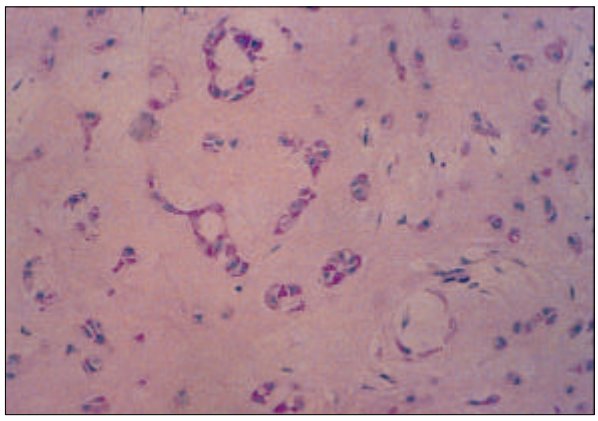

Fig. 4. The tubular alveolar structures contain PAS positive diastase resistant amorphous eosinophilic materials (PAS, × 400).

DISCUSSION

Headington JT5 histologically classified chondroid syringoma into two types: those tumors with tubular, cystic, partially-branching lumina and those tumors with small, tubular lumina. Chondroid syringomas with tubular, branching lumina show marked variation in the size and shape of the tubular lumina, and they also show cystic dilatation and branching.

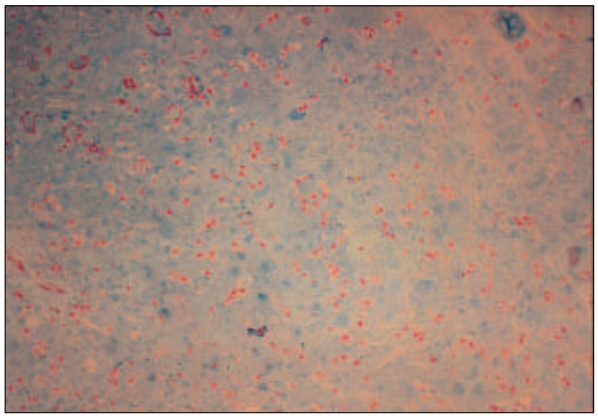

Fig. 5. The chondroid matrix shows a faintly bluish mucinous substance (Alcian blue, × 200).

The tubular lumina are embedded in an abundant stroma, and they are lined by two layers of epithelial cells: a luminal layer of cuboidal cells and a peripheral layer of flattened cells. On the other hand, those chondroid syringomas with small, tubular lumina show small ducts, as well as small groups of epithelial cells and solitary epithelial cells scattered through a mucoid stroma. The tubular lumina are lined by only a single layer of flat epithelial cells, and from these lumina, small, comma-like proliferations often extend into the stroma, and these resemble syringoma. Our case is consistent with this small tubular lumina type.

Headington JT5 also recognized that chondroid syringomas with apocrine origin demonstrated irregular branching tubules lined by at least 2-cell-thick epithelum, and tumors with eccrine origin demonstrated rather uniform, small, round tubules lined by a single cell layer of epithelial cells. He suggested the architecture of the latter should be compared with that of eccrine spiradenoma due to the striking similarity.

Klein W et al.[3] insisted that tumors with tubular branching lumina were much more common than those with small tubular lumina. This finding correlated with those findings of Headington JT5 and others reports[7,8]. In the same manner, among the seven cases reported in the Korean literature, only one case[6] corresponded to the small tubular lumina type.

Although most of these reported cases have been benign, a few cases of malignant chondroid syringoma have been described as having a different clinical presentation than the relatively usual benign forms. These malignant forms generally present as a larger lesion and they are more likely to occur on the trunk and extremities of females[9]. Histologically, the presence of celluar atypia, increased mitotic activity, infiltrative margins, tumor necrosis and satellite tumor nodules are considered to be indicative of malignant behavior[9,10]. Infiltration of the regional lymph nodes and metastasis to the bones and visceral organs has been reported. In most of these cases, anaplastic changes were present from the beginning. Only rarely does a chondroid syringoma of many years suddenly undergo a malignant transformation[11].

The treatment of choice is complete excision, along with a cuff of normal tissue, in order to examine the histopathologic features, and recurrence does not occur unless residue tumor has been left unexcised.

The previously reported incidence of chondroid syringoma among all the skin lesions was less than 0.01%, whereas in a large series reported recently by Yavuzer R. et al.[4] the incidence was detected as 0.098%, which is far higher. Hence, the report[4] suggested that although they are rare lesions, for differential diagnosis of small subcutaneous nodules in the head and neck region of middle-aged male patients, chodroid syringoma must be remembered, and these lesions must be completely excised to confirm a definitive diagnosis, without jeopardizing the important aesthetic component and functional status of the patient.

We herein report on a rare case of chondroid syringoma with small tubular lumina that occurred on the scalp of a 67-year-old Korean woman.

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