A 72-year-old man with a mass in his right axilla

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Case report

The patient was a 72-year-old man with a mass in his right axilla for 1 year. The tumor was painless, mobile, and slowly increasing in size; at its largest it measured 3 cm in diameter and was in a subcutaneous location. The clinical presumptive diagnosis was epidermoid cyst, mucocele, adnexial tumor, and nodule. The lesion was excised under local anesthesia.

The resected specimen consisted of well-defined round and slightly fluctuant dermal nodules. Its size was 3 × 2.5 × 2 cm. The operative specimen proved to be completely cystic, and the cyst contents contained brownish serous fluid (Fig. 1). The thickness of the cyst wall was at most 0.2 cm.

Histological examination revealed the absence of an epithelial lining in some areas and in some cells with apocrine-type decapitation secretion along the inner surface of the cyst wall (Fig. 2). The section showed a tumor composed of epithelial cells and connective tissue stroma, the two being intimately admixed (Fig. 3). The epithelial pattern was tubular with a double layer of lining cells, branching strands cords, and focally solid areas. Most of the stroma was of dense myxoid appearance and stained positively with PAS. Immunohistochemically, the inner cell layer of tubular structures was positive for cytokeratine, epithelial membrane antigen, and the cells of the outer cell layer were positive for vimentine and S-100.

K E Y W O R D S

chondroid syringoma, cystic, axilla

What is your diagnosis?

Totally cystic chondroid syringoma of the axilla.

Discussion

A mixed tumor, a term described by Minssen in 1874, is characterized by the coexistence of epithelial and mesenchymal features (1). Chondroid syringoma (CS) was first suggested as a name for the mixed tumor of the skin by Hirsch and Helwig because the lesion is epithelial and has merely secondary changes in the stroma (2). Many other names have been used, including syringomatous adenoma, eccrine syringofibroadenoma, adnexal tumor, and eccrine sweat-duct tumor (3). Tumors that arise from salivary glands are more commonly referred to as pleomorphic adenomas, whereas those that arise from sweat glands are referred to as CS, or mixed tumors of the skin (4). CS represents a dermal neoplasm of epithelial/myoepithelial differentiation (5). Tumors composed almost entirely of myoepithelial cells without obvious epithelial ductal differentiation are referred to as myoepithelioma (1). The incidence of CS has been reported between 0.01 and 0.098% among the primary skin tumors (2, 6). CS is usually a firm intracutaneous or subcutaneous nodule, but sometimes focal cystic degeneration occurs (6). To our knowledge, total cystic degeneration of CS has not been reported, although two cases of parotid benign pleo-

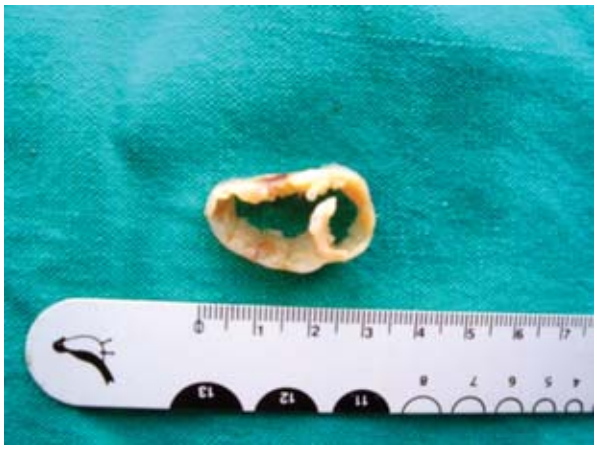

Figure 1.Cystic tumor from the axilla.

morphic adenoma with extensive cystic degeneration have been reported (7).

CS is relatively rare benign cutaneous appendiceal tumor that usually affects males and arises in the dermis and subcutaneous fat of the head and neck region; other locations are the axilla, trunk, and extremities (8). In 80% of cases it is observed in elderly patients (6). The male to female ratio is 2:1. Clinically, CS is usually a small, painful, nonulcerated, solitary, benign, slowly growing skin tumor (2, 9). It characteristically presents as a firm intracutaneous or subcutaneous nodule. The lesion is commonly movable and separated from surrounding tissues. The size is usually between 0.5 and 3 cm, but it may be very large (2, 10).

Clinical recognition is difficult and a histopathological confirmation is required. CS can be diagnosed by fine needle biopsy, which is recommended prior to surgery (11). The differential diagnosis includes epidermoid cysts, mucoceles, adnexial tumors, dermoid implantation, sebaceous cysts, compound nevus, clear cell hidradenoma, cystic basal cell carcinoma, neurofibroma, dermatofibroma, or a granulamatous process (4, 11). The tumor is usually benign, but malignant transformation and metastases have been rarely reported (4, 11). A local recurrence is possible and follow-up is suggested (13).

Neither the origin of the glandular component nor which cell is responsible for the production of the unique myxoid stroma is known. It is hypothesized that the cells may arise from an abnormal outgrowth of an ectodermal analage or cell rest that has maintained its pluripotent nature. Results from ultrastructural studies have suggested that the plasmocytoid cells found in the stroma exhibit myoepithelial differentiation and therefore may represent the cell responsible for the production of the myxochondroid stroma (4). Other authors have suggested that the myoepithelial cell plays only a minor role in the development of mixed tumors and that it is filamenteous cells, which are derived from the ectodermal analage, that produce the myxochondroid stroma (4).

CS has been sub-classified as apocrine or eccrine. Approximately 80% of cases are apocrine; eccrine CS is uncommon (12). Histopathologically, it is similar to pleomorphic adenoma of the salivary glands (in the presence of a complex admixture of epithelial derived structures arranged in ductal-glandular formations, tubuloalveolar structures, branching patterns, and solid areas, as well as in the stroma, which is variably condromyxoid to hyalinized (8)). Histologically the tumors are classified into two types. Type 1 is the more common variant, it is lined by double layered epithelium exhibiting decapitation secretion characteristic of apocrine glands. Type 2 also contains ducts with small lumina lined by only a single-layered epithelium (4, 13). Immunohistochemically, the inner cell layer of tubular structures expresses cytokeratine and epithelial membrane antigen, whereas the outer cell layer expresses vimentine and S-100, as in our case. Chondroid stroma stains positively with PAS, but both with

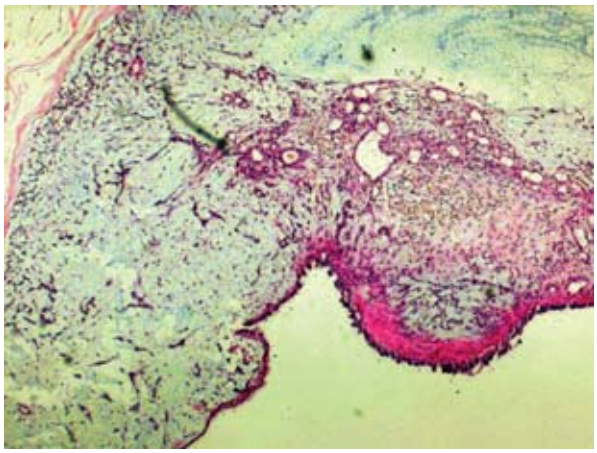

Figure 2. Luminal surface of cystic space and deeper portion of the wall (H&E, original magnification × 10).

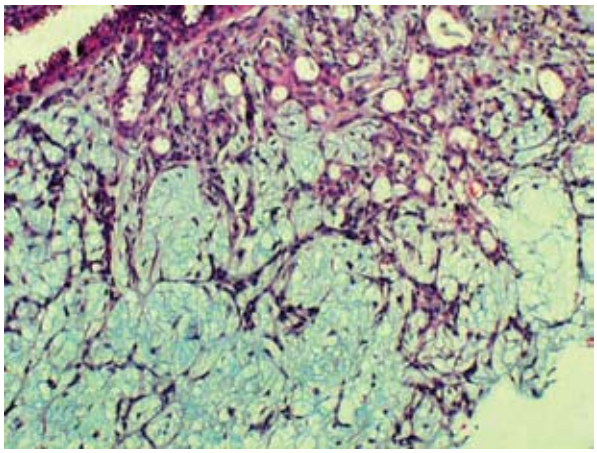

Figure 3. Cords and tubules of epithelial cells in a myxoid stroma (H&E, original magnification × 20).

and without diastase digestion; it also satins positively with mucocarmine and Alcian blue at both low and high PH (4). Complete resection of the primary tumor is recommended. The diagnosis can be usually made with hematoxylin and eosin staining, but immunohistochemical methods may be sometimes necessary.

A totally or partial cystic degeneration, tiny hemorrhagic foci, and small infarcted areas are occasionally seen in benign pleomorphic adenoma of parotis, which strongly resembles CS (7). A focal adipocytic metaplasia, hemorrhage, and cholesterol deposition may occur (5, 6), but we did not observe extensive cystic degeneration.

In conclusion, CS is a rare epithelial tumor. We reported a case of totally cystic, benign CS in the axilla; this form of CS has not been described in the literature. In our case, totally cystic degeneration is most likely due to a spontaneous change in the lesion. The diagnosis of the tumor as solid or focal cystic is clinically difficult. The total cystic form should be included with benign and malignant subcutaneous tumors.

Learning points

- Chondroid syringoma, or mixed tumor of the skin, represents a benign dermal or dermal and subcutaneous neoplasm of epithelial/myoepithelial differentiation with a typical chondroid stroma;
- Histologically similar tumors arising from salivary glands are more commonly referred to as pleomorphic adenomas;
- Complete resection of the tumor is curative.

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