MALIGNANT CHONDROID SYRINGOMA IN THE ANTERIOR ABDOMINAL WALL: A RARE CASE REPORT

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Abstract: Malignant chondroid syringomas (MCS), which are also called malignant mixed tumors of the skin, are extremely uncommon tumor arising from the eccrine sweat glands with tumor differentiation in the epithelial and mesenchymal tissues. Malignant chondroid syringomas commonly involve the limbs, head and neck rarely trunk. In this article, we present a case of malignant chondroid syringoma localized in the anterior abdominal wall. Wide local excision was followed by close observation. After 3 years of close observation the patient remains asymptomatic. MCS is rare with 43 reported cases in the literature. The site of the primary tumor was the lower extremity in 35 percent, the head in 28 percent, and the upper extremity in 23 percent. As MCS may progress very slowly and disease recurrence including metastasis occurs in a relatively high percentage of cases, long-term follow-up of MCS cases is required.

Introduction: Chondroid syringomas (CSs) are rare, benign tumors of the skin. In 1859, Billroth first described them as a mixed tumor of the skin due to its histopathological resemblance to the benign mixed tumor originating from salivary gland. [1, 2] The term "chondroid syringoma" in lieu of mixed tumor of the skin was introduced by Hirsch and Helwig in 1961. They proposed the following microscopic diagnostic criteria: (a) Nests of cuboidal or polygonal cells; (b) Intercommunicating tubuloalveolar structures lined with two or more rows of cuboidal cells; (c) Ductal structures composed of one or two rows of cuboidal cells; (d) Occasional keratinous cysts; (e) Matrix of varying composition in hematoxylin and eosin stain. [3] In 1961, Headington divided CS into two groups, including apocrine type and eccrine type, based on their histopathological appearance. [4] Chondroid syringomas most commonly occur in the head and neck with a size ranging from 2 mm to ≥1 cm and usually present with solitary, solid, painless, nonulcerative, subcutaneous, or intracutaneous nodule. [5, 6] They often affect middle-age to elderly patients with a male-to-female ratio of 2: 1. [6-8] The malignant variant of CS is rare. In most cases of MCS, anaplastic changes are present from the beginning. Malignant chondroid syringoma is a mixed cutaneous tumor, with epithelial and mesenchymal components, which compromises principally the trunk and extremities. Rarely, a chondroid syringoma of many years duration suddenly undergoes malignant changes with widespread metastasis. [9] There have been no reports reporting effectiveness of chemotherapy and radiotherapy, and an early wide excision with a broad margin may be the most reliable treatment to date. In this paper, we present a case of MCS localized in anterior abdominal wall.

Case Report: A 55-year-old female was admitted in 2010 at the Department of Surgery, Bankura Sammilani Medical College with a slowly growing nodular lesion at the anterior abdominal wall. The lesion was a painless, slow growing nodular lesion gradually increasing in size for last five years. On
物理检查，一个孤立的，半球形，皮肤颜色的，坚实的结节，测量35 mm x 35 mm，被注意到在前腹部壁5 cm上方和侧方至腹直肌。肿瘤没有粘附到腹直肌鞘。从病变的FNAC结果是不可信的。结节被手术切除，并且1 cm的边缘被移动。术后皮肤缺损被关闭。手术标本显示肿瘤细胞的腺样增生与少数的有丝分裂细胞，嵌入在粘液组织中（图1）。免疫组织化学显示肿瘤细胞的大多数是PAS消化酶， toluidine blue， cytokeratin (CAM5.2, CK7)， S-100蛋白，和GCDFP-15。从上述发现，诊断为软骨样腺瘤是成立的。手术切除的边缘是阴性的。区域淋巴结的超声波检查和胸部和腹部的X光片显示没有证据的 metastasis。该患者，经手术后三年保持无病，仍然每三个月进行随访。


metastases and distant metastases were observed in 39% and 36% of the cases, respectively. The most common site for distance metastasis was lung, followed by bone and brain. In conclusion, negative surgical margin should be attained using wide excision technique. These patients should be also monitored closely due to high potential of recurrence.

CONCLUSIONS: Malignant chondroid syringoma tends to appear between 4th to 5th decades of life and follow an unpredictable clinical course. The neoplasm has a preponderance to affect limbs mostly (61%), while its benign counterpart affect mainly in the head and neck (80%). Diagnosis was based on the wide surgical excision of the tumour. Combination chemotherapy in patients with metastasis is not reported to be beneficial. In conclusion, negative surgical margin should be attained using wide excision technique. These patients should be also monitored closely due to high potential of recurrence.

REFERENCES:
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LEGEND: Fig 1: Histopathological examination of excised mass. Tumour showed a lobular proliferation of tumour cells with glandular differentiation associated with anaplastic changes and mitotic cells, in the background of a mucinous stroma. In addition, generalized lymph vascular invasion was present (HE, ×200).

Fig. 1

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