



## **METASTATIC MALIGN CYLINDROMA; CASE REPORT.**

### **Metastatik malign silendroma; Olgu sunumu.**

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#### **ABSTRACT**

Cylindroma is a rare form of slow-growing dermal neoplasm which appears as painless, smooth, pink and skin coloured, solitary or multiple nodules. These tumors are most commonly seen over scalp or neck. Malignant transformation of cylindroma is unusual and extremely rare in solitary type. Surgical removal of growth is of primary importance. Prolonged careful follow-up and screening of family members is essential in the management of this disease. We report a case of malignant cylindroma of scalp with lung and bone metastases.

**Key words:** Cylindroma, malign, and turban tumor.

#### **ÖZET**

Silendroma cildde renk değişikliği, ağrısız, düzgün sınırlı, tek veya multipl lezyonlar şeklinde kendini gösteren, nadir görülen ve yavaş büyüyen bir tümördür. Genellikle saçlı deri ve boyunda görülürler. Soliter lezyonların malignite potansiyeli kazanması son derece nadirdir. Lezyonun cerrahi olarak çıkarılması en önemli tedavidir. Aile bireylerinin kontrolü erken tanı ve tedavide önemlidir. Burada, akciğer ve kemik metastazlı malign silendromalı bir vakamızı sunduk.

**Anahtar kelimeler:** Silindroma, malign, türban tümörü.

#### **INTRODUCTION**

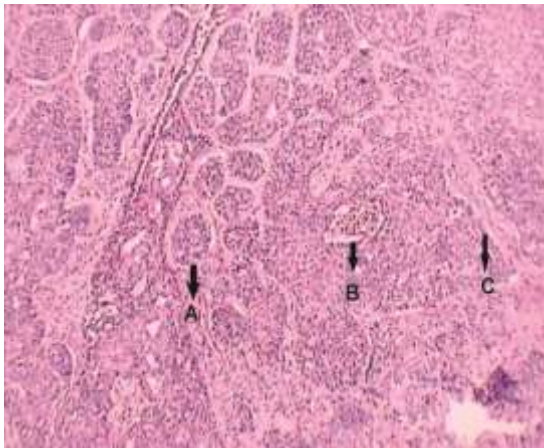
Cylindromas are uncommon benign skin appendages tumor that are most frequently found in scalp and neck skin, with a strong predilection for middle-aged and elderly females (1). Malignant cylindroma was first recognized by Ancell in 1842 in a family, the members of which presented with multiple scalp tumors (2). Billroth and Beobachtung coined the term cylindroma in 1859 due to the epithelial elements of such growth being enclosed in well-defined 'cylinders' of connective tissue (3). These tumours are of two distinct clinical types, solitary and multiple(1,4). Solitary cylindromas occur most commonly on the skin of the head and neck and occur sporadically and typically are not inherited (1,5). Multiple cylindromas are inherited in an autosomal dominant pattern and occur on the scalp and rarely on the trunk and extremities (1,6). Multip-

le lesion can cover the entire scalp giving the appearance of a turban, hence the eponym turban tumor (6). In 1929, Widemann first described malignant cylindroma (7). Malignant transformation of a solitary cylindroma is extremely rare whereas it is unusual in patients with multiple lesions (1).

#### **Case**

A 35 year old woman presented with complaint of a large painful ulcerative turban-like growth over the most part of the scalp 15 years back. There was accompanying intermittent bleeding and itching. Growth was initially a cluster of small, painless, nodular lesions spreading over the upper portion of the scalp. There was no family history of such type of lesion. On a routine checkup, X-Ray chest revealed cannon ball shadow in the right lung. CT guided FNAB was planned from right lung le-

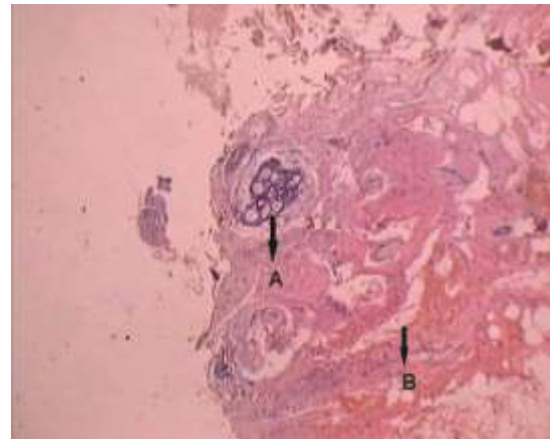
sion and showed metastatic deposit from malignant cylindroma.



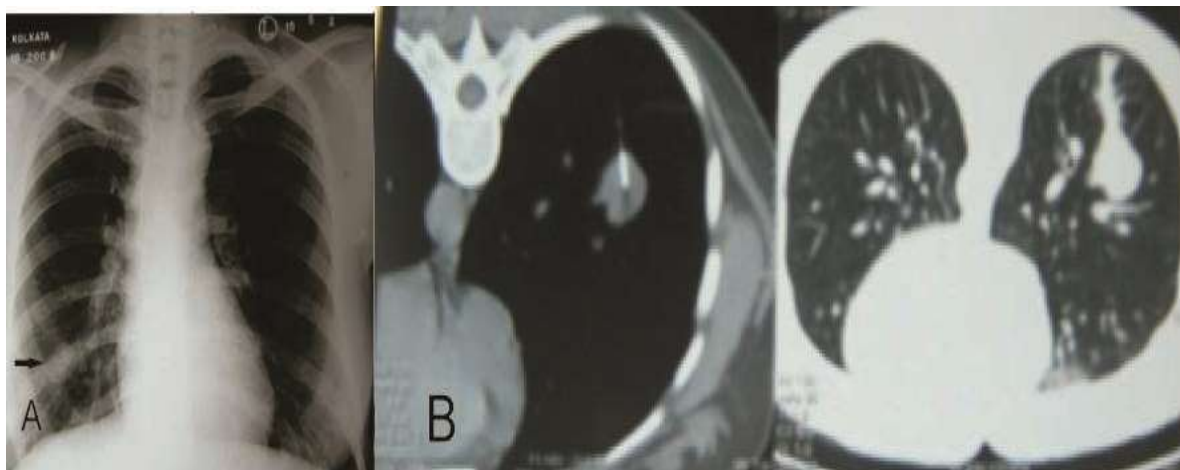
**Figure 1:** Malignant cylindroma showing compressed benign tumor showing tubular pattern (A) cellular pleomorphism, high mitotic count and absence of jig-saw pattern (B) strands of hyaline tissue (C) (Haematoxyline & Eosine stain x 40).

Extensive excision of scalp tumor followed by plastic reconstructive surgery was done and histopathologically malignant cylindroma was confirmed from excised tissue. She also received adjuvant combination chemotherapy (cisplatin, doxorubicin and cyclophosphamide) and external beam radiotherapy over chest following surgery. One year after surgical removal of primary tumor, a nodular growth appeared again over the scalp. This growth was surgically removed followed by reconstruction and histopathologically found to be recurrence of malig-

nant cylindroma. Patient was kept under close observation and she remained almost asymptomatic for next four years. Four months prior to this reporting, nodules started rapid growth with appearance of pain, itching, ulceration and occasional bleeding. She also complained of gradually increasing left sided chest pain, mostly on the left scapular region radiating towards anterior chest along with respiratory difficulty. Radiograph of chest showed massive pleural effusion on left thorax. Whole body <sup>99m</sup>Tc-MDP bone scan revealed multiple metastatic lesions in the skeleton.



**Figure 2:** Recurrent Malignant cylindroma shows glandular spaces surrounded by malignant cells (A) Soft tissue of scalp (B) (Haematoxyline & Eosine stain x 40) .



**Figure 3:** Chest X-ray shows right sided coin shaped opacity (A) and CT scan shows nodular lesion right lower lobe (B).

### DISCUSSION

Cylindromas are benign skin appendage tumors with two distinct clinical presentations, namely, solitary and multiple (1,4). Solitary cylindromas occur most commonly on the skin of the head and neck and occur sporadically and typically

are not inherited (1,5). Malignant transformation of a solitary cylindroma is extremely rare and is somewhat more common, but unusual (33 cases reported) in patients with multiple lesions (4). The histological features of malignant changes are loss of hyaline sheets, prominent large cells, nuclear anaplasia and

pleomorphism, stromal invasion, loss of jigsaw puzzle arrangement, focal areas of necrosis, squamous cell metaplasia, and loss of palisading at the tumor island periphery (8). Metastases may be found in regional lymph nodes, stomach, thyroid, liver, lung, and bones. In our case metastases was seen in multiple bones in addition to pulmonary involvement. Treatment modalities are simple excision, scalping, Mohs' micrographic surgery, laser ablation.

In conclusion; malignant cylindroma is a rare aggressive tumor with tendency of local destructive growth and potential to metastasize. Therefore, early diagnosis offers the patient the best options for management. Long term careful follow up and screening of family members are of prime importance in the management of such neoplasm.

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