Case Report

Askin tumor in an adult female

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ABSTRACT

Askin tumor is extremely rare and difficult to diagnose. We present a 55-year-old female with multiple pleural and diaphragm based masses on the right side. Contrast enhanced computed tomography chest showed multiple pleural and diaphragm based masses on the right side. Fine-needle aspiration cytology and Plural biopsy were done. Both studies proved it to be Askin tumor. Immunohistochemistry for cluster of differentiation 99 was positive. This case to our knowledge is very rare as the patient is elderly, and the previously reported cases of Askin tumor were male. Here, we discuss the dilemma in clinical and pathological diagnosis of such a rare case with previously reported cases in the literature.

Key words: Askin tumor, cluster of differentiation, contrast enhanced computed tomography chest, fine needle aspiration cytology, plural biopsy

INTRODUCTION

Primitive (or peripheral) neuroectodermal tumor (PNET) and Ewing sarcoma (ES) both are malignant, small, round-cell tumors of bone and soft tissue.[1] With the advent of immunohistochemical, cytogenetic, and molecular genetic techniques, it is almost universally regarded that these tumors represent ends of a morphologic spectrum known as the ES/PNET family of tumor.[2] Identification of a common cytogenetic abnormality, t (11;22) (q24;q12), in ES and PNET clearly supports the contention that these neoplasms are histogenetically related.[2]

In 1979, Askin *et al*. described the "malignant small cell tumor of the thoracopulmonary region" as having histologic features similar to those of PNET but with a unique clinicopathologic profile.[2,3] Although not a different entity ES/PNET located in the chest wall have been defined as Askin tumors.

In our case, we report 55-year-old female with Askin tumor. Askin tumor is a disease of adolescents or young adults, the majority of whom are <30 years of age, and there is a slight male predilection.[2] In older age group, we found only two reported case of Askin tumor presented at age of 67 years and 73 years (both male).[4,5]

We report here one further case of Askin tumor occurred in older age (female).

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CASE REPORT

A 55-year-old female with no history of smoking was admitted to hospital for chest pain cough on September 2012. She was suffering from these symptoms for the last 3 months. Chest X-ray showed a right-sided mass with right-sided pleural effusion.

Contrast enhanced computed tomography chest confirmed multiple pleural and diaphragm based masses on the right side measuring 112.6 mm × 71 mm × 70.4 mm, 42.9 mm × 41.9 mm and 10.8 mm × 9.3 mm [Figure **1**]. With the right-sided pleural effusion with lytic lesion in D_3 vertebral body. Other laboratory tests including pleural fluid examination were within normal limits.

Fine-needle aspiration cytology (FNAC) showed – blood mixed cellular aspirate with occasional clusters and few groups of loosely cohesive intermediate sized tumor cells showing high nuclear (N):cytoplasmic (C) ratio, nuclear molding, overlapping, coarse reticular–granular chromatin and small to inconspicuous nucleoli; cytoplasm is scant, light basophilic. Tumor cells showed

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trabecular arrangement with formation of numerous rosettes and few mitotic figures [Figure 2]. A diagnosis of - Malignant small blue round cell tumor (Askin tumor/ PNET) was given.

Pleural biopsy showed – sheets of small to intermediate sized round tumor cells infiltrating into the fibrocollagenous stroma. The tumor cells showed trabecular arrangement, occasional rosette formation, high N: C ratio, nuclear molding, inconspicuous nucleoli and scant eosinophilic cytoplasm [Figure 3]. A diagnosis of Malignant small round blue cell tumor, suggestive of Askin tumor was given.

Immunohistochemistry (IHC) was positive for cluster of differentiation 99.

DISCUSSION

Askin tumor is a subset of ES that arises from the chest wall. It is characterized histologically by features of small round blue cell tumors. It was described by Askin *et al*. in 1979.[3] Adolescents or young adults are mainly affected, majority of whom are <33 years of age; mean age of 18 years.[6]

The gross appearance of the tumor varies. In general, it is multilobulated, soft, and friable; it rarely exceeds 10 cm in greatest diameter. The cut surface has a gray-yellow or gray-tan appearance, often with large areas of necrosis, cyst formation, or hemorrhage. Despite the extensive necrosis, calcification is rare.[2]

The histologic features of typical Askin tumor is composed of sheets or lobules of small round cells containing darkly staining, round or oval nuclei. The cytoplasm is indistinct except in areas where the cells are more mature, and the elongated hair-like cytoplasmic extensions coalesce to form rosettes. Most of the rosettes are similar to those seen in neuroblastomas and contain a central solid core of neurofibrillary material (Homer Wright rosette). Rarely, the rosettes resemble those of retinoblastoma and contain a central lumen or vesicle (Flexner-Wintersteiner rosette). Some tumors are composed of cords or trabeculae of small round cells. These areas bear a resemblance to a carcinoid tumor or a small cell undifferentiated carcinoma, although histogenetically they are properly compared with the primitive neuroepithelium.[2]

Askin tumor must be differentiated from other small round cell tumors like - rhabdomyosarcoma, neuroblastoma, desmoplastic small round cell tumor, mesenchymal chondrosarcoma, lymphoblastic lymphoma, etc.[2]

Until the introduction of modern therapy only a small percentage of patients with this tumor survived.

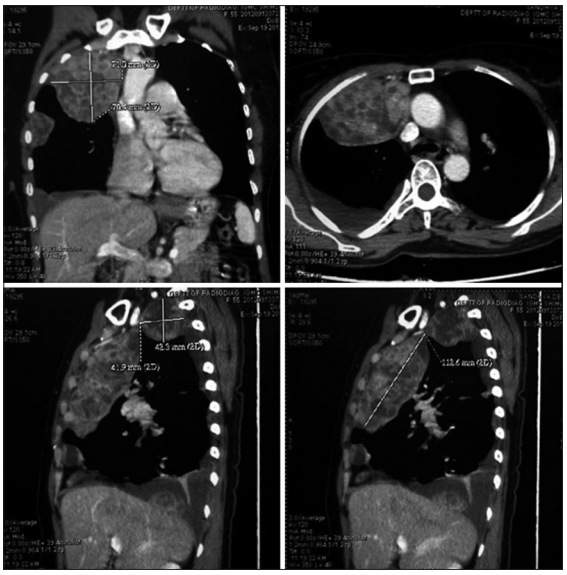

Figure 1: Contrast enhanced computed tomography chest shows multiple pleural and diaphragm based masses on right side

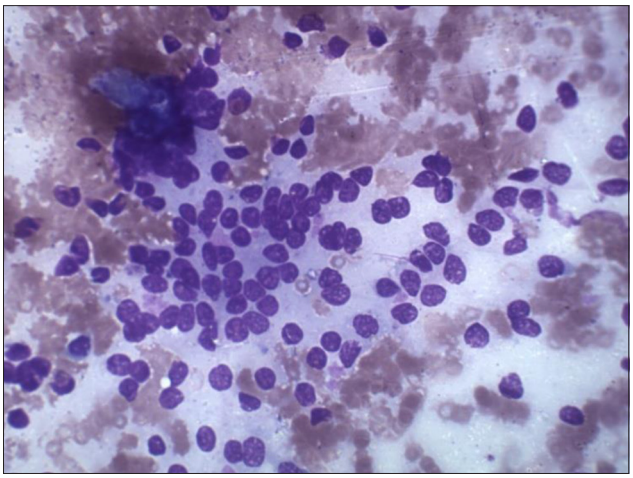

Figure 2: Fine needle aspiration cytology of tumor cells showed trabecular arrangement with formation of numerous rosettes

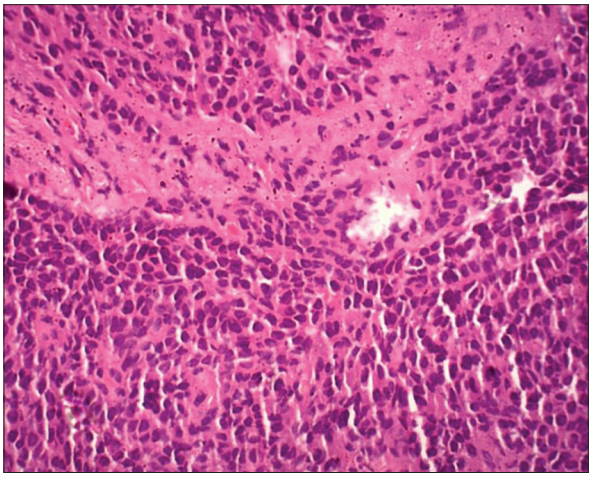

Figure 3: The tumor cells showed trabecular arrangement, occasional rosette formation, high nuclear: cytoplasmic ratio, nuclear moulding, inconspicuous nucleoli and scant eosinophilic cytoplasm

The prognosis for patients with Askin tumor has steadily improved. About 75% of patients present with localized disease, and the combination of surgery and/or radiotherapy and systemic chemotherapy results in a cure rate near 75% in this group. Krasin *et al*. reported 90% survival at 10 years for patients with localized disease treated with surgery and multiagent chemotherapy.[2] Yuki Nakajima *et al*. reported a case of Askin tumor who has survived for 10 years after two occurrences of lung metastasis; 5 years and 9 years after initial surgery and post-operative radiotherapy.[7] However, patients with metastatic disease at presentation have a long-term cure rate of <30%, even with high-dose chemotherapy, followed by stem cell reinfusion. Key prognostic factors that adversely influence the outcome of the disease are the presence of metastatic disease at the time of initial diagnosis, large tumor size, extensive necrosis (filigree pattern), central axis tumors, and poor response to initial chemotherapy.[2]

CONCLUSION

Askin's tumor is a rare tumor of mainly young adults. However, it may be seen in old age also. It would require a high index of suspicion in any chest wall mass in patients. Image-guided pleural biopsy and IHC is helpful for diagnosis. However, considering its aggressive nature, complicated course and recurrence tendency, long-term followup is warranted.

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