Pulmonary involvement in malignant glomus tumor

Sir,

Glomus tumors are uncommon lesions that usually occur as solitary lesions at dermis or subcutis area of the upper or lower extremity with the most common site at the subungual regions of fingers.[1] These are almost exclusively benign in nature with malignant lesions extremely uncommon and metastasis is further rare. Pulmonary involvement either as primary or secondary involvement is again very rare.[2] This report describes a case of young adult with malignant glomus tumor in the leg with recurrence and subsequently pulmonary involvement as a large metastatic lesion.

A 22-year-old male presented with a chief complaint of progressive breathlessness, dry cough, and right-sided chest pain for the last 4 months. He was nonsmoker and driver by occupation.

He gave a history of gradually increasing painful swelling on medial aspect of leg above medial malleolus 1 year back for which he was fully investigated. Magnetic resonance imaging leg report revealed a heterogeneous well-defined mass of 16 cm × 3.8 cm posterior to the tibia and anterior to the tendoachilles with a possibility of neurogenic tumor [Figure 1a] and complete excision was done. The histopathological report showed features of soft tissue small round cell tumor with possibility of Ewing's sarcoma or peripheral primitive neuroectodermal tumor [Figure 1b] and further immunohistochemistry markers were positive for Vimentin, actin, smooth muscle actin, CD34, and IN-I. The lesion showed focal positivity to pan-cytokeratin (CK), TLE-1, and ki-67 and negative for desmin, BCL-2, S-100, ERG, HMB-45, CD-99, EMA, and CD-45. Based on the above findings, he was diagnosed as a case of malignant glomus tumor of the leg. He subsequently received adjuvant chemotherapy and radiotherapy as per protocol. However, even after starting of therapy, there was two times recurrence of painful swelling at the same site for which again excisions were done with similar histological findings. He then received adjuvant radiotherapy, but could not complete it due to grade 4 skin toxicity. A chest radiograph and computed tomography scan after repeat surgery for persistent cough revealed a round well-defined opacity at the right upper lobe [Figure 2a and b] that could not be investigated further at that time. He was referred to us for the same reason.

On examination, surgical scar was present over the medial aspect of left ankle. There were no clubbing or palpable lymph nodes. On respiratory system examination, there was dull percussion note and reduced intensity of breath sounds over the right infraclavicular and mammary area with no adventitious sounds.

Routine investigation showed normal blood counts, liver and renal function test, etc. A recent chest X-ray [Figure 3] revealed a large homogenous opacity with well-defined margins at the right upper and middle zone suggestive of rapid enlargement of previously detected pulmonary lesion. A guided fine-needle aspiration and subsequent percutaneous transthoracic core biopsy from the lung mass revealed clusters of small-sized cells having little or no cytoplasm with pleomorphism and high N/C ratio. The cells showed hyperchromatic nuclei with granular nuclear chromatin and numerous mitotic figures suggestive of metastatic deposits of small round cell carcinoma [Figure 4]. A final diagnosis of metastatic glomus tumor at the lung was made based on correlation and review with previous biopsy tissue histology examination. The patient was referred to medical oncologist for further management.

Glomus tumors are relatively uncommon benign mesenchymal lesions with an estimated incidence of approximately 1.6% among the soft tissue tumors.[3] These are distinct neoplasms composed of perivascular cells that resemble modified smooth muscle cells of the normal glomus body. The latter is a specialized form of arteriovenous anastomosis that serves in thermal regulation and is located in the stratum reticularis of the dermis and is most frequently located at the subungual region, lateral areas of the digits, and palm. The glomus tumor is therefore common at these sites; however, the tumor can also be encountered rarely at other sites where the normal glomus body is sparse or even absent,

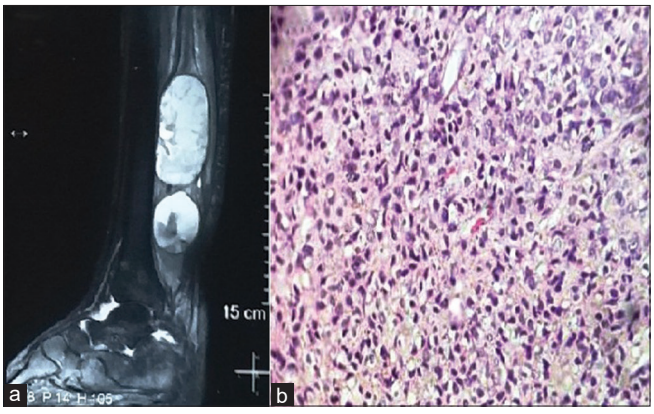

Figure 1: (a) Magnetic resonance imaging leg showing a large soft tissue mass at lower part of leg. (b) Photomicrograph of excised mass showing sheets of uniform round cells with intervening blood vessels, hyperchromatic cells with bland chromatin and mitoses (H and E, ×40)

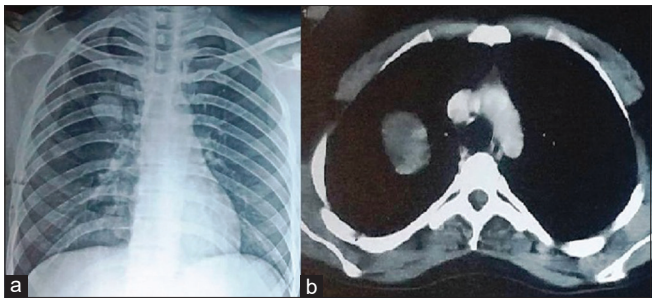

Figure 2: X-ray chest (a) and computed tomography chest (b) showing a well-defined rounded opacity at right upper lobe

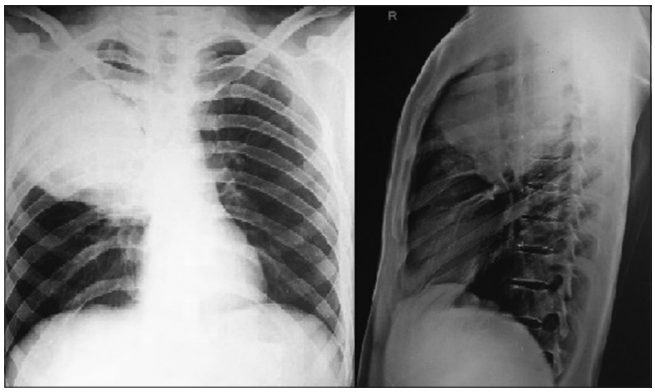

Figure 3: X-ray chest PA and right lateral view showing a large mass lesion at the right upper zone and rapid increase in lesion size compared to Figure 2

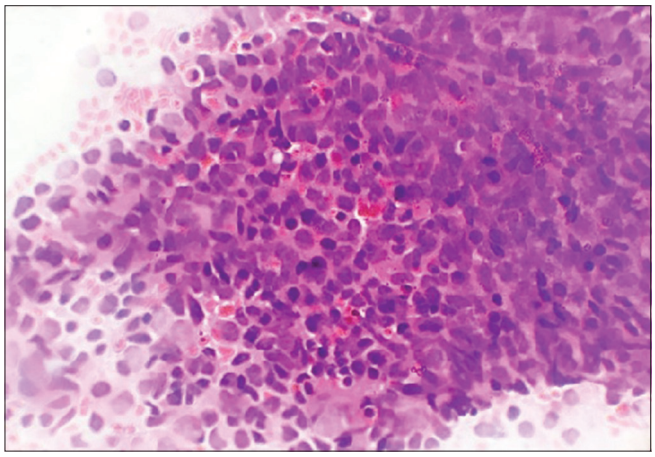

Figure 4: Photomicrograph of fine-needle aspiration smear from lung mass showing features of metastatic deposits of malignant glomus tumor (H and E, ×40)

such as mediastinum, respiratory tract, gastrointestinal tract, gynecological regions, etc.[4]

These tumors are usually solitary and well circumscribed, but multiple lesions may also occur. They tend to be asymptomatic and usually acquired during the childhood. Almost all of these tumors are benign both histologically and clinically; however, malignant glomus tumors are exceedingly rare and their diagnosis is based on histological criteria.[5] Malignant tumors demonstrate aggressive clinical and histological features, such as nuclear atypia, infiltrative growth pattern, and multicentricity. Pulmonary glomus tumors are very rare and mostly benign with malignant lesions further rare either as primary or secondary involvement. Malignant glomus tumor may be seen from childhood to elderly with male predominance.[6] Clinical symptoms in pulmonary tumors are nonspecific such as cough, chest pain, and hemoptysis as seen in our case. They may present as endobronchial lesion as well to cause postobstructive consolidation.

The differential diagnoses of malignant glomus tumors or glomangiosarcoma at lung include carcinoid tumor, hemangiopericytoma, epithelioid leiomyoma, paraganglioma, non-Hodgkin lymphoma, melanoma, adenoid cystic carcinoma, and primitive neuroectodermal tumors. Histological and immunohistochemical features help differentiate them. Glomus tumors generally stain negative for keratin 18, CK, chromogranin A, CD34, S-100 protein, and synaptophysin, but positive for actin, desmin, Vimentin, epidermal growth factor receptor, and excision repair cross-complementation.[7]

Benign tumors usually respond to local excision. Malignant glomus tumor behaves aggressively with tendency to recurrence and distant metastasis as seen in our case. They resemble high-grade sarcomas in clinical course. There is no definitive recommended therapy for aggressive metastatic malignant glomus tumors although palliative therapy and chemotherapy remains option in such lesions. Clinical evaluation, careful histological examination, and immunostaining with appropriate markers are essential to confirm the diagnosis of malignant glomus tumors. Currently, it is difficult to accurately predict metastasis on clinical ground and prognosis in malignant glomus tumors with pulmonary involvement. Therefore, close follow-up of such patients is warranted.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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