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Case Report

Primary Osteosarcoma of Pulmonary Artery - A Case Report and Review of Literature

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Abstract

Primary sarcomas of pulmonary artery are extremely rare, highly malignant tumours and primary osteosarcoma of pulmonary artery is even rarer. Most of the cases are often misdiagnosed as pulmonary thromboembolism and anticoagulated to no effect. We present a case of 38-year-old male initially treated for infection and pulmonary thromboembolism but did not respond to antibiotics and anticoagulative measures. Chest radiograph revealed multiple lung nodules. Excision of right main pulmonary artery mass and upper lobe lung nodules was performed and was proven to be an osteogenic sarcoma of primary pulmonary artery on histopathological examination.

Keywords: Osteosarcoma; Pulmonary Artery; Sarcoma

Introduction

Primary tumours of the pulmonary arteries are rare and most of the reported cases are sarcomas [9]. Primary osteogenic sarcoma of pulmonary artery is extremely rare. Clinical presentation is of pulmonary thromboembolism refractory to anticoagulation and this frequently leads to a delayed diagnosis [1]. A case of primary osteosarcoma of pulmonary artery with predominant chondroblastic component in this case report is presented.

Case Report

A 38-year-old gentleman presented with fever, cough, chest pain and dyspnea on effort for 2 months, unresponsive to treatment and sought medical advice from various hospitals. He was initially treated for infection with antibiotics. On subsequent examination, multiple nodules were noted on chest radiograph and pulmonary thromboembolism was suspected on chest computed tomography. The patient was anticoagulated to no effect and possibility of neoplasm was suggested. Right main pulmonary artery mass and upper lobe nodules were excised and submitted for histopathological examination. An initial diagnosis of primary intimal artery sarcoma with metastasis to the lung was made. Subsequently, the tissue was referred to this centre for second opinion.

The tumour was composed of malignant spindle shaped cells forming osteoid and chondroid elements. Herring bone and storiform pattern of spindle cell tumour were also observed. The tumour cells showed hyperchromatic or vesicular nuclei with prominent nucleoli. Mitoses were seen. Immunohistochemical studies showed heterogeneous positivity for osteocalcin. A final diagnosis of primary pulmonary osteosarcoma of pulmonary artery with predominant chondroblastic component and metastases to the lung was made.

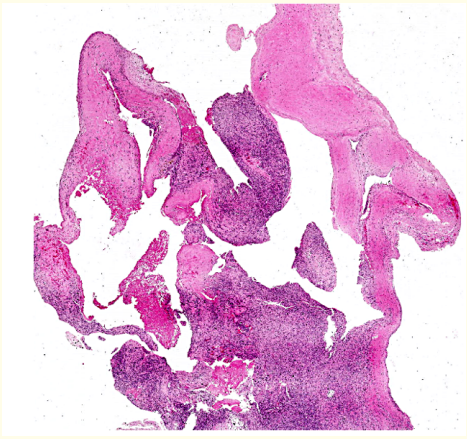

Figure 1: Low power view - Cellular tumour @ magnification of 2x

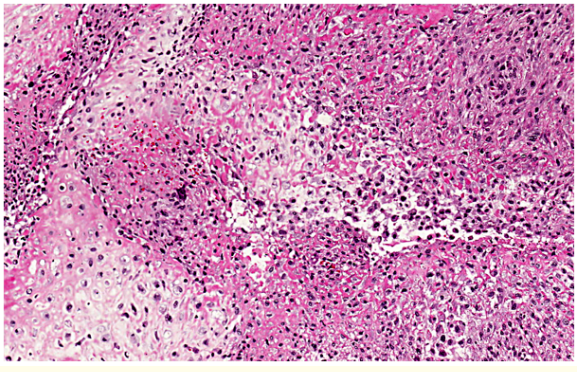

Figure 2: Osteosarcoma with malignant chondroid and osteoid components at magnification of 10x

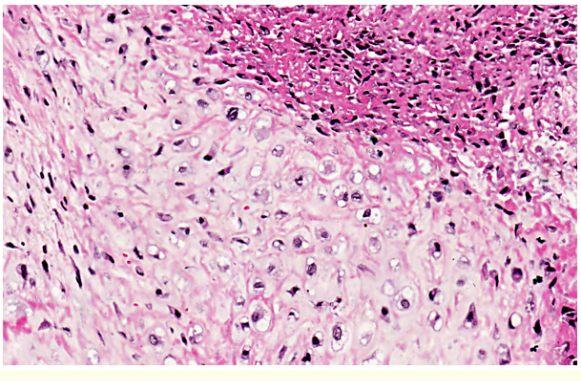

Figure 3: Malignant chondroblastic component @ magnification of 20x

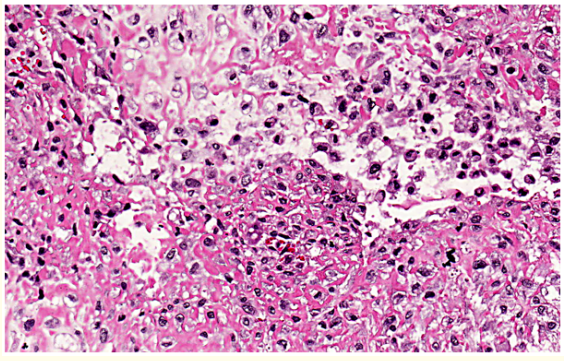

Figure 4: Lace like osteoid at a magnification 0f 40x

Discussion

The first case of primary sarcoma of the pulmonary artery was reported from an autopsy by Mandelstamm in 1923 [2]. Ramp., *et al*. reported that approximately 60% of cases of primary pulmonary artery sarcoma were diagnosed at autopsy [3]. More than two hundred cases are reported in the literature but many are underestimated through misdiagnosis as pulmonary embolism, supported by clinical and radiological findings [4-6]. Clinical symptoms of pulmonary artery sarcomas are nonspecific such as dyspnoea, chest pain and cough [1]. Clinical features that can aid in the differentiating the tumours from pulmonary embolism include fever, elevated erythrocyte sedimentation rate, anaemia, weight loss, absence of procoagulant state and particularly lack of history of deep vein thrombosis [7,9]. Most cases are initially being treated for thromboembolism before a suspicion of tumour is made [8]. Due to recent advances in imaging technology, appropriate use and interpretation of different imaging studies can significantly facilitate accurate diagnosis [1,4,9]. Blackmon., *et al*. proposed a staging system for primary pulmonary artery sarcomas to determine optimal therapy for management of these patients [7].

Pathological classification of primary sarcoma of the pulmonary artery is based on the histological subtype and location of the tumour (luminal versus mural) [9,10,32]. Most sarcomas of the pulmonary artery arise from the intimal layer and can show wide variety of histological differentiation [9]. The variable differentiation could be due to origin of pulmonary artery sarcomas from the undifferentiated tissues of the bulbous cordis as suggested by Bleisch., *et al*. [11,12]. However, no significant correlation between histological type of the tumour and prognosis has been identified [1,7,13].

Osteogenic sarcoma of the pulmonary artery is an extremely rare form of intravascular tumour. Osteosarcomas usually arise from the metaphyseal ends of long bones; however, extraosseous cases have been reported in soft tissues [19,20], retroperitoneum [21], lung [17], heart [14-16], uterus [18] and thyroid gland [22]. The wall of the pulmonary artery is the common site for osteosarcoma similar to that of sarcomas of pulmonary artery [23]. Histopathological examination confirms the diagnosis of osteosarcoma. The presence of chondroblastic component may contribute to better prognosis in some of these patients [17,29]. To the best of our knowledge, ten case reports of primary pulmonary artery osteosarcoma have been reported [13,23-31]. Pneumonectomy was done in three cases. Pulmonary embolectomy was done in two cases and one of them had pulmonary arteriotomy and extraction of tumour done. The rest of the cases were diagnosed at autopsy. Mc Connell reported the longest survival in his case for 51 months following recurrence at 9 months of initial diagnosis [24]. Cases reported by Madu., *et al.* and Kimura., *et al.* died 5 days and 7 months respectively following surgery [23,27]. The outcome of the case reported by Lyerly., *et al*. was unknown [28]. The patient was alive for 2 years in the case report published by Tsunezuka., *et al*. The recent case published by Forbes., *et al*. died three days after operation due to multiorgan failure [31]. The prognosis is generally poor and the recurrence rate is high. Despite complete surgical resection of the tumour, distant metastasis and recurrences can occur. Hence, a careful follow up is mandatory in these cases [29].

Conclusion

Primary osteogenic sarcoma of pulmonary artery frequently masquerades as thromboembolic disease. Due to its rare occurrence and potential limitations of imaging to rule out a neoplasm, a high index of clinical suspicion should be borne in mind especially in cases unresponsive to anticoagulant therapy. Clinicians should be aware of this disease and has to consider in the differential diagnosis when investigating cases of pulmonary artery obstruction.

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