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Neurology

UNUSUAL CLINICAL SCENARIO – SPINDLE CELL TUMOR PRESENTING AS CARDIO-EMBOLIC STROKE

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ABSTRACT We report a case of 39-year-old female, known case of left thigh spindle cell sarcoma, who presented with right cerebellar and right fronto-parietal subcortical infarct within the window period. IV tPA was administered, following which, patient had developed left hemothorax. On resolution of hemothorax, patient was found to have metastatic lesions in both lung fields. On further imaging of chest, metastasis was found to invade the pulmonary vein and entering into left atrium. We report a case of unusual and rare cause of cardio-embolic stroke in patient with spindle cell sarcoma.

KEYWORDS : Spindle cell sarcoma, Cardio-embolic stroke, Metastasis.

INTRODUCTION

Spindle cell sarcoma is a rare subtype, constituting 2% of soft tissue sarcomas (1). It can arise from any anatomical site, however more than half of such tumours originate within an extremity (2). It can develop from a variety of reasons including genetic susceptibility, inflammation, injury etc. Such lesions are highly metastatic, especially to lungs, pleura and abdomen. Spindle cell sarcomas arising out of extremities have high predilection for metastasis. Thrombo-embolic events in this disease are exceedingly rare. Tumour embolization is one of the rare causes of embolic infarcts. Majority of literature for tumour embolization is available for atrial myxoma. Here in this case report we are presenting an aggressive spindle cell sarcoma causing pulmonary vein invasion extending into left atrium, acting as a source of embolization.

CASE REPORT

A 39-year-old female had medical history of left thigh high grade spindle cell sarcoma and had received chemotherapy and neo-adjuvant therapy, presented to the emergency of other hospital with abrupt onset left sided weakness since 3hrs. Examination at admission revealed a conscious, alert patient with left hemiplegia. Both pupils were 2mm symmetrical with normal reaction to light. Motor power in left upper limb and left lower limb was 0/5. Left plantar response was extensor. Patient was afebrile with pulse rate of 90/min and blood pressure of 150/70 mmHg. Multiple ulcerated lesions seen over antero-lateral aspect of left thigh. NIHSS at admission -14/42. Urgent MRI head performed which revealed acute infarct in right cerebellum and right subcortical fronto-parietal region (Figure. 1). Patient meeting all the indications for iv thrombolysis, hence IV tPA was given with dose of 0.9 mg/kg. After 30 minutes of ongoing thrombolysis, patient had developed sudden hypotension and drowsiness.

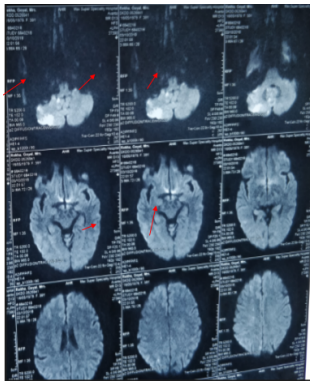

Figure 1MRI head showing acute infarct in right cerebellum and right subcortical fronto-parietal region

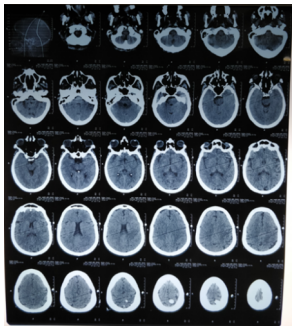

Figure 2- NCCT head after aborting thrombolysis

IV tPA infusion was aborted and urgent NCCT head was done (Figure. 2) . There was no bleeding from the wound on left thigh. NCCT head didn't reveal any haemorrhage, subsequently patient was shifted to ICU and was intubated in view desaturation. Chest X ray revealed left hemithorax opacity, which was found to be hemothorax and ICD was inserted and subsequently patient was shifted to our centre for further evaluation. Patient underwent CECT Chest for evaluation of underlying metastasis, which revealed bilateral nodular lesions, left lesion was extending into pulmonary veins and left atrium (Figure 3). 2D Echo revealed similar mass into openings of pulmonary veins in left atrium. Left sided chest lesion was biopsied twice, which revealed spindle cell sarcoma (Figure 4). Pleural fluid analysis didn't reveal any malignant cells. Patient was managed with statins, packed red cell transfusion, dressing of the wound on left thigh and other supportive care.

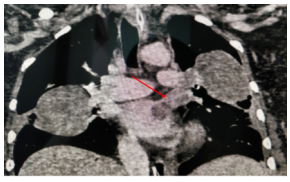

Figure 3- CECT Chest revealed bilateral nodular lesions, left lesion was extending into pulmonary veins and left atrium

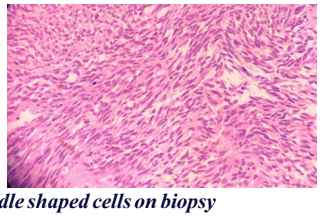

Figure 4- Spindle shaped cells on biopsy

DISCUSSION

Acute phase therapy in ischemic stroke includes IV tPA within window period of 4.5 hours (3,4). Incidence of symptomatic ICH after IV tPA is 6-8 %, found in many studies. Spindle cell sarcomas, being quite rare subtype of soft tissue sarcomas, have been seen to have high metastatic potential, especially to lungs, pleura and abdomen. Spindle cell sarcoma is classified as an undifferentiated sarcoma. It is characterized by lack of specific histology with negative immunohistochemistry for desmin and keratin. Such undifferentiated sarcomas typically involve the pulmonary vessels, left atrium and may extend to the pericardium resulting in a hemopericardium.

Complete surgical resection remains the only possibility for cure from pulmonary metastases in soft tissue sarcoma; however, only 11% of patients with an extremity sarcoma whose first distant recurrence is in the lung will have a survival of around 3 years, despite therapy. Complete resection and the development of more effective adjuvant treatments are imperative to improve outcome for this group of patients (1). For the management of cardio-embolic stroke, anti-coagulation is warranted. Choice of anti-coagulant is based on whether the atrial fibrillation is valvular or non- valvular. There are no recognized clinical management guidelines for managing these patients owing to the rarity of cases and undetermined pathophysiology for the cardio-embolic events. As the embolus contains tumour cells, role of anti-platelets and anticoagulants are doubtful, rather can be detrimental due to risk of bleeding.

The treatment for soft tissue sarcoma is surgical removal, cryotherapy, radiotherapy, hormone therapy and chemotherapy. They may also be combined together for best results. If the cancer has undergone metastasis, the best treatment options are chemotherapy and radiotherapy. However, the chance for survival is very low (5).

CONCLUSION-

This case emphasises the importance of careful staging and characterization of these tumours including cardiac and cerebral MRI and screening for metastasis, so that surgical planning and treatment decisions may be tailored for individual patients by a multidisciplinary approach.

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