A 58-year-old male was admitted to our hospital with symptoms of numbness, paraesthesia, and mild weakness of the right forearm. Gadolinium-enhanced magnetic resonance cerebral imaging showed a large mass in the right precentral region with strong contrast enhancement (Panel A). This alteration was suspect for metastasis of an unknown primary tumour. Subsequently performed computed tomography (CT) of the thorax demonstrated partial occlusion of the aortic arch by a lobulated intraluminal mass (Panels B and C) as well as total, non-calcified occlusion of the left common carotid artery. Computed tomography scan also revealed a suspicious tumour of the left adrenal gland and left-sided second rib (not shown). To further characterize, the pathology of the aortic arch transesophageal echocardiography was performed. Hereby an irregularly shaped, partial mobile mass originating from the aortic wall could be visualized in the aortic arch and descending aorta (Panels D and E).

Computed tomography-guided needle biopsy of the second rib was performed. Histologic findings revealed undifferentiated pleomorphic tumour cells showing atypic mitosis figures and hyperchromatic nuclei (Panel F). In summary histologic, CT and echocardiographic findings strongly suggested an undifferentiated aortic intimal sarcoma.

Since the tumour already had multiple satellites curative surgery was not an option. The patient was treated with palliative chemotherapy (ifosfamide and doxxyrubicine) and received cerebral radiation (30 Gy). He died 3 months later due to fast progress of his metastatic malignancy.

Primary aortic sarcoma is a rare and usually fatal malignant disease typically presenting with neurological symptoms due to early cerebral metastasis or malperfusion signs following central or peripheral vessel occlusion.