East African Medical Journal Vol. 92 No. 1 January 2015 SMOULDERING ONSET OF ADVANCED MEDIASTINAL TUMOR IN AN ADOLESCENT BOY: CASE REPORT G. Gaido, MD, DTM&H and C. Lanza, MD, PhD, Cottolengo Mission Hospital, Chaaria, Meru, Kenya

Request for reprints to: Giuseppe Gaido, MD. Medical Director, Chaaria Cottolengo Mission Hospital, Meru, Kenya. Email: fr.beppe@gmail.com

SMOULDERING ONSET OF ADVANCED MEDIASTINAL TUMOR IN AN ADOLESCENT BOY: CASE REPORT

G. GAIDO and C. LANZA

SUMMARY

An adolescent boy came at our observation with bilateral flaccid paralysis at both legs, cough, fecal and urinary incontinence. First symptoms started 6 months before, were initially mild and rather aspecific. There was incomplete information available from former diagnostic imaging and histologic assessment. We describe and discuss the subsequent diagnostic flow driving to the final diagnosis of spindle cell mediastinal carcinoma, its clinical management and outcome. Hopefully, sharing our lesson learnt may rise awareness around this rare tumor in adolescent population, in order to better anticipate diagnosis and improve its outcome.

INTRODUCTION

A 17-year-old boy was referred to Chaaria Mission Hospital with a six months history of gradual bilateral flaccid paralysis at lower limbs, associated with anaesthesia of the same area and urinary / fecal incontinence. He didn't report any trauma and felt otherwise healthy before the onset of paralysis. The young patient started complaining of cough since one month before admission.

A previous MRI of the spine performed in a referral hospital, showed signal alteration on the body of T9 vertebra with pathological tissue invading the vertebral body, left peduncle, lamina and neuroforamen, and compressing the spinal cord epidurally, without significant contrast enhancement. Following this finding, in August 2013, the boy underwent neurosurgery attempt on the dorsal spine (probably excisional biopsy, but no documentation was available).

CASE PRESENTATION

When the boy was admitted in our hospital in December 2013, he was febrile (T 38.8°C) but overall stable. He presented with associated anaemia (the patient had never been transfused) and diaphoresis, with particularly extensive sweating at night. On examination we observed left lung hypophonesis with reduced ventilation and non productive cough. The neurological examination confirmed a complete

flaccid paralysis at both the legs, and associated sphincter incontinence.

Additional investigations included an unremarkable Pelvis X-Ray, Chest X-Ray with features of left extensive pleural effusion, Chest U/S (which we performed in absence of CT scan locally available) showing extensive leftlung atelectasia, though pleural effusion was not evident.Full Blood Count revealed moderate anemia (Hb 6.6 g/dl), WBC 9.800/ml (granulocytes 77%, lymphocytes 14%).HIV test was negative. ESR was increased 104 mm/h (partly due to anemia). Liver and renal function tests, electrolytes were all within normal ranges.

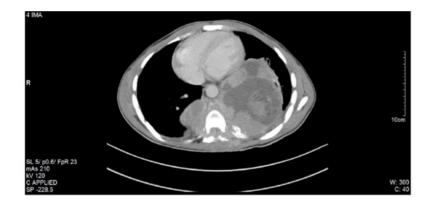
There was also a deep sacral bed sore, evident on admission, hence the patient was treated with a myocutaneous flap from the gluteus, with partial improvement. The patient received physiotherapy immediately after admission to our hospital, but unfortunately without improvement of the lower limb paralysis.

In January 2014 we empirically decided to start him on anti-TB treatment (sputum could not be obtained for microbiological confirmation), together with large spectrum antibiotics, considering the clinical history, with associated involvement of the left lung and the MRI imaging with T9 infiltration (suggesting a possible TB of the spine).

However when the neuroradiological documentation was sent for a second opinion, it was suggested also a possible neoplastic origin of the lesion at T9. There was indeed evidence of a mass at the level of T9, at the same level of the previous surgery, but expanding and extending from T5 to T12. The mass was oval shaped, approximately 4 cm wide, looking fleshy and not containing bone tissue. Considering the rapid increase in size of the dorsal mass during the hospitalisation, we decided to perform an open biopsy (previouslya fine needle aspirate was also done but it was not helpful, as showing skeletal muscle and foci of mild non specific inflammation). The biopsy sample was then sent for histology review to a reference lab, which identified "tissue exhibiting proliferation of spindle cells on an inflamed and necrotic background". Spindle cells showed mild atypia, and mitoses not easily identified. PAS and ZN stain were negative, in line with the diagnosis of Spindle Cell Neoplasm. The biopsy procedure was followed by subsequent progressive discharge of necrotic material from the wound, which resulted in reduction of the neoplastic mass.

Based on the histopathology report we decided to stage the neoplastic disease. We referred the patient to Nairobi for a CT scan which revealed few subcentimeter metastatic nodules in the right lung. The scan also showed a 15 x 10 cm mass in the posterior mediastinum with lobulated margins (Figure 1), which was described as invasive and extensive. The mass was also displacing the aorta anteriorly and invading the left pericardium, the posterior lung segments and the left superior lung segment. The mass was also causing erosion of the T9 vertebra and anterior angulation of the spinal column. There was extension of the mass inside the spinal canal from T7 to T9 causing severe cord compression. Adjacent ribs were showing osteolytic lesions and the mass was also crossing the left diaphragm inferiorly.

Figure 1 *Legend.* Chest CT scan showing tumor invasion in the left lung, pericardium and spinal canal



An abdominal CT scan did not show any metastatic localisation.

The advanced stage shown by imaging, confirming metastatic localisation and infiltration of the spinal canal, appeared clearly beyond any reasonable possibility of curative intent, other than palliative care for advanced cancer. We immediately stopped the anti-TB treatment because unnecessary at that point and potentially hepatotoxic for the patient.We started our patient with intra venous steroids, which significantly improved the dyspnoea and his appetite. We continued with nursing care and mobilisation in order to prevent worsening of bed sores. After seven month the patient unfortunately progressed, both locally with the development of a permanent fistula discharging necrotic, foul smelling material; and systemically with general deterioration, extreme anorexia, cachexia and extension of the bed sores to all the pressure points. The boy eventually passed away last September .

DISCUSSION

The main learning from this case is around the reasons delaying the primary diagnosis, due to the ambiguous and progressive onset of the disease. Within several potential causes of posterior mediastinal masses, including both oncology and non-oncology indications (1, 2) the histology of spindle cell neoplasms is not the most expected in a boy with a large mass spreading and expanding in the posterior mediastinum after involving vertebral bodies and spinal cord. Shading some light on this case and sharing our lesson learnt may hopefully

raise further the awareness and understanding on these rare cancers, and help anticipate the diagnosis of similar patients, in order to improve chances for earlier optimal management and better outcome.

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