An unusual case of focal epilepsy

BMAC Balasuriya¹, Jagath C Wijesekara² and Sunil Perera³

(Index words: Cerebral aneurysm, embolisation, myxoma)

Introduction

Primary tumours of the heart are rare. Three fourths of these tumours are benign, and nearly half of these benign tumours are myxomas. Embolisation from left atrial myxoma to the cerebral hemispheres, brainstem, cerebellum, retina and spinal cord has been recorded. Embolisation may take the form of a transient ischaemic attack or infarction. We discuss here a case of multiple cerebral metastatic deposits of a cardiac myxoma, which presented as focal fits several years after resection of the primary tumour.

Case report

A 30-year old mother of two children was admitted to the Neurology Unit at National Hospital of Sri Lanka, on 2 January 2004 with a history of persistent weakness of the right arm following an epileptic fit 1 week ago. She gave a history of similar attacks for 2 years. Her fits started with a tingling sensation involving fingers of the right hand, that gradually spread to involve the whole of her right arm. Despite antiepileptic medication she was experiencing infrequent attacks of fits.

Her past medical history revealed hospitalisation for resection of a cardiac myxoma in the year 2000. Trans-oesophageal echocardiogram after surgery showed no residual myxoma tissue in the cardiac chambers, and she was discharged from the clinic. She had an uncomplicated pregnancy 1 year later. Though the first attack of fits was observed during the first trimester, up to her delivery the fits were infrequent. She was lost to follow up for the next 2 years.

Clinical examination at our unit in January 2004 revealed a thin woman, weighing 42 kg, with no pallor or facial pigmentation, thyroid nodules, lymphadenopathy, or ankle swelling. Pulse rate was 76/min and BP 130/80 mmHg, apex beat not shifted, there were no thrills and auscultation revealed no murmurs. Respiratory and abdominal examinations were normal. Central nervous system examination showed normal higher functions, with mild congestion of the right optic disc. Her visual acuity was normal, and all other cranial nerves were normal. Muscle tone in her right arm and leg was slightly increased, power was Grade 4– in the right arm and grade 4+ in the right leg. Plantar reflex was extensor on the right side.

Investigations

Hematological investigations including Hb, MCV, MCHC, WBC/DC and thrombophilic screen were normal, with ESR 28 mm 1st hour, Blood urea 6 mmol/L, Na 138 mmol/L, K 3.8 mmol/L; Ca 2.56 mmol/L 2D echocardiogram ECG and chest radiograph were normal. Trans-oesophageal echocardiogram showed normal cardiac chambers devoid of myxoma deposits. Ultrasound of abdomen was normal. CT scan of the brain revealed multiple hyperdense lesions involving both cerebral hemispheres (Figure 1).

White matter oedema was marked on the left side. Ventricles appeared normal. The appearances were compatible with multiple metastatic deposits from the primary thyroid, malignancy, choriocarcinoma, or melanoma. A malignancy screening was carried out, including fine needle aspiration cytology of a thyroid nodule. Urine HCG was negative. She had no evidence of melanoma. VDRL and HIV tests were negative. As the CT scan appearances were inconclusive, and clinical findings were unremarkable, save for the fact that she had mild weakness of right arm, a tissue diagnosis was considered mandatory.

A stereotactic brain biopsy was performed. The histology showed tumour composed of stellate cells, and multinucleated cells in a abundant mucoid matrix, which stained strongly with Alcian blue, with evidence of haemorrhagic areas, compatible with myxoma tissue. MRI scan of the brain was compatible with multiple cerebral secondaries. The magnetic resonance angiography failed
to show flow void areas within the lesion, suggesting aneurysm formation.

**Discussion**

In this case the diagnostic problem was that the patient’s medical records of the year 2000, after total resection of the primary cardiac tumour, revealed no evidence of residual myxoma tissue in the heart, albeit her cranial CT demonstrated multiple metastatic deposits widely scattered in both cerebral hemispheres. Two possible postulates can be offered to explain this phenomenon. Firstly, the embolisation of this myxoma would have taken place at the time of original resection of the tumour, and the deposits in the cerebral tissue would have grown ever since. Secondly, the myxoma that embolised at that time must have lodged in the cerebral vasculature, forming a mycotic aneurysm, which ruptured some years later releasing the contents, and the local invasive property of the myxoma led to the florid lesions. Although the magnetic resonance angiography failed to show aneurysm formation, the possibility cannot be ruled out without performing a digital subtraction angiography.

Emboli occur at a frequency of 30–50% with myxoma. Left atrial myxoma gives rise to systemic embolisation. In the majority of cases cerebral arteries are involved. Transient or permanent visual loss may result from involvement of retinal arteries. The behaviour of embolised tumour fragments, within the central nervous system is controversial. In rare instances tumour fragments continue to grow and thus create a symptom complex, compatible with an expanding intracranial mass [5,3]. A rarer delayed complication of embolised myxoma is arterial aneurysm at the site of embolisation [1,2]. These aneurysms enlarge progressively and can cause neurological symptoms, years after the removal of the primary tumour [4]. Rupture of such aneurysms has been reported [1,2,5]. Angiographic studies have shown aneurysmal change in cerebral vasculature [2]. It has been postulated that a tumour embolus weakens the arterial wall and permits viable myxoma cells to penetrate the damaged endothelium, causing weakening of the media, and aneurysmal dilatation. A cerebral arteriogram may demonstrate this as a filling defect.

As this patient had evidence of multiple lesions, a surgical removal was not feasible. Our patient was managed with intravenous steroids to reduce the cerebral oedema and antiepileptic medication to control the fits.

**References**


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**An unusual presentation of female urethral leiomyoma**

**Neville D Perera¹, Lakshman Senanayake⁴, V Hasanthi Vithana² and Ruchira Fernando³**

(Index words: Diagnostic approach, histology, urethral and bladder reconstruction)

**Abstract**

Leiomyoma of the urethra is a rare clinical entity. We report an unusual presentation of this tumour, which led to a diagnostic and surgical dilemma. The patient was a 16-year old female who presented with a labial mass which was palpable abdominally. Imaging methods and laparoscopy demonstrated a well defined soft tissue mass arising from the pelvis, without any obvious involvement of the urinary tract. The tumour (8 × 10 cm) was completely excised with reconstruction of the bladder and urethra. Histology confirmed a cellular leiomyoma.

**Introduction**

Urethral leiomyoma is rare and only about 50 cases have been reported in literature. This benign smooth muscle tumour often presents as a midline extramucosal vaginal mass, which is often mistaken for a urethral diverticulum, fibroid or prolapse. We report an unusual presentation of a large lateral tumour, which led to a dilemma in diagnosis and surgical approach.

¹Urological Surgeon, ²Research Assistant, Department of Urology, and ³Pathologist, Department of Pathology, National Hospital of Sri Lanka; ⁴Obstetrician and Gynaecologist, Castle Street Hospital (Teaching) for Women, Colombo. Correspondence: NDP, e-mail: <nevi603@sltnet.lk> (Competing interests: none declared). Received 15 June 2004 and accepted 20 August 2004.