DOI: https://doi.org/10.47391/JPMA.704

An unusual presentation of Ewing sarcoma mimicking Guillain–Barré syndrome in a 17-year-old male

Ozgur Deniz Sadioglu[1], Ozgur Sogut[2], Barıs Agca[3]

1,2 Department of Emergency Medicine, University of Health Sciences, Haseki Research and Training Hospital, Istanbul, Turkey; **3** Department of Neurology, University of Health Sciences, Dr.Ersin Arslan Training and Research Hospital, Gaziantep, Turkey

Correspondence: Ozgur Sogut. **Email**: ozgur.sogut@sbu.edu.tr

Abstract

The management of a patient admitted to the emergency department with symptoms of Guillain–Barré syndrome (GBS), including paraplegia, who was subsequently diagnosed with Ewing sarcoma (ES) and spinal cord compression using MRI is discussed here. Pathological report confirmed the diagnosis of ES. The patient underwent immediate neurosurgery due to rapid progression of paraplegia.

Keywords: Ewing sarcoma, Paraplegia, Guillain–Barré syndrome, Spinal cord compression

Introduction

Guillain-Barré syndrome (GBS) is a demyelinating polyneuropathy of the peripheral nervous system in which the body's immune system attacks its own nerves. GBS is characterised by rapid-onset weakness and areflexia in the lower extremities that may progress to the arms, face, and oropharyngeal muscles.[1, 2] Sensory deficits associated with GBS are symmetric and distal.[2, 3] Ewing sarcoma (ES) is a member of the Ewing family of small, round-cell tumours in

children and young adults. ES is the second-most common malignant bone tumour after osteosarcoma.[4] The majority of cases develop in patients aged five to 25 years, most frequently between the ages of 10 and 20 years. Metastasis is present in the initial stage of the disease in one out of four cases.[5]

Here, we describe the management of a patient admitted to our emergency department (ED) with GBS symptoms, including paraplegia, who was subsequently diagnosed with ES and spinal cord compression using imaging techniques.

Case

A 17-year-old Turkish man was admitted to the ED of our tertiary-care university hospital (University of health sciences, Haseki training and research hospital, Istanbul, Turkey) in October 2018. Five days earlier he had developed ascending numbness, starting in the toes, and loss of strength. He stated that he had mild headache that began six months ago after minor head trauma. The patient did not lose consciousness at any point. On examination, his orientation and cooperation were good, his Glasgow coma scale (GKS) score was 15/15, his pupils were isocoric and IR +/+, eye movements were normal, but partial ptosis of the left upper eyelid was noted. Bilateral upper extremity muscle had strength and was fully functional; however, the bilateral lower extremities were plegic from T10 down and showed numbness and areflexia. Deep-tendon reflexes were positive (+/+) in the upper limbs and absent (-/-) in the lower limbs. The base skin reflex was weak bilaterally. Urine retention was detected, and lumbar puncture revealed albuminocytological dissociation. The patient was screened for GBS in the clinic because ptosis of left upper eyelid suggested cranial nerve involvement. Brain computed tomography revealed an 8.5-mm-thick hyperdense area in the epidural space at the extra-axial distance adjacent to the right frontal lobe (Figure-1). Epidural haemorrhage was excluded due to

the absence of a history of head trauma or evidence of bleeding on brain magnetic resonance imaging (MRI).

MRI of the posterior bone structures at C2–3, the spinal canal at T1–2, spinal canal, and posterior bone structures and spinal cord extending to about 23 mm revealed a 38-mm mass in the spinal canal surrounding the T6–8 vertebral corpus. Multiple mass lesions, scattered diffusely around the bony structure of the L3–4 vertebrae, were considered primary metastases because of the intense contrast enhancement and large number of lesions (Figure-2). A 10 x× 9-mm lytic lesion was detected in the proximal one-third of the left femur (Figure-3).

The patient underwent immediate neurosurgery because of rapid progression in paraplegia. Emergency decompression of the T6-8 vertebrae was performed to rescue the spinal cord from compression. Macroscopic examination of 8 cc of excised irregular tissue fragments revealed a small round, blue tumour cell. However, histochemical and immunohistochemical findings (positive staining with CD 99) were suggestive of ES/primitive neuroectodermal tumour. Preoperative chemotherapy consisting of Vincristine, Doxorubicin and Cyclophosphamide was given and 45 Gy radiation was administered as local treatment. Postoperative care was uneventful. Two weeks after the surgery, the patient was transferred to the rehabilitation department. Approximately six weeks after the surgery, motor power in the lower limbs improved to grade III-IV, and the patient was able to walk a short distance with assistance. At the postoperative six-month follow-up, the motor power had improved to grade IV-V for both legs. He could walk about freely without any support.

Discussion

Axial tumours at ES are located primarily in the pelvis and chest wall, vertebra and paravertebral fields, or the skull. Circulating ES cells are found in 30% of patients with localised disease and in 50% of those with metastatic disease. Furthermore, metastasis develops in 10% of patients with classical ES and in

35% of those with atypical ES and primitive neuroectodermal tumour.[4] The tumour may compress the spinal cord or peripheral nerves, depending on the effect of the primary mass or metastasis. Spinal cord compression is a rare oncological emergency in children (4.0–5.5%) and is the most frequent cause of paraparesis in individuals between the ages of three months and 17 years.[6] Masses arising from the spinal cord or channel (astrocytoma, and ependymoma) are primary, whereas solid masses are secondary tumours that cause spinal cord compression.[7] In children, secondary tumours are the most common cause of spinal cord compression. In adults, spinal cord compression is frequently caused by metastasis to the vertebral corpus, whereas in children, spinal cord compression is generally the result of paravertebral tumour extension into the intervertebral space.[5, 7] ES may cause spinal cord compression via direct contact with the vertebrae or paravertebral field. The pressure of the tumour on the vertebral venous plexus stimulates the release of inflammatory mediators, which cause vasogenic oedema, venous bleeding, and ischaemia, resulting in neurological deficits and tissue damage.[4, 8] The clinical findings in our case, including the absence of muscle strength in lower extremities on neurological examination, areflexia, and the presence of sensory deficits made us consider a diagnosis of spinal cord compression.

Emergency diagnosis and treatment are critical when neurological symptoms are present because long-term compression of the spinal cord may cause irreversible neurological damage. At diagnosis, 80–95% of patients have pain. Motor disorders (60–85%) and sensory loss (40–90%) occur in the days following. Urinary incontinence or constipation and urinary retention may occur as a result of autonomic dysfunction.[8, 9] Physical examination typically reveals an increase in deep tendon reflexes and Babinski sign. Muscle strength, anal sphincter tonus, and deep and superficial sensory examination are important for the assessment of spinal cord dysfunction.[9] Although our patient reported pain, it was mild and was thought to be due to the trauma experienced six months

earlier; moreover, the major complaint at the current visit to our hospital was motor and sensory deficits in the lower extremities.

Direct radiography has proven a useful diagnostic tool for detecting osteolytic or osteoblastic changes in the structure of the vertebrae.[7] MRI is the most effective modality for imaging the spinal canal and the epidural and paravertebral areas; the technique is non-invasive, has high specificity and sensitivity, and provides multiplane, cross-sectional images. If MRI cannot be performed, computed tomography, positron emission tomography, and radionuclide bone scintigraphy are alternative imaging methods.[8-10] In our case, MRI revealed a mass and spinal canal compression; thus, no further imaging studies were necessary. Our aim was to establish the presence of GBS and to determine whether intracranial lesions were the cause of the ptosis.

For masses of unknown cause on the spinal medulla, surgery is the preferred treatment to achieve spinal decompression and obtain diagnostic information. Other treatment options for maintaining neurological function in patients with acute spinal cord compression include Dexamethasone, radiotherapy, decompression surgery, and chemotherapy.[8, 11] Corticosteroids are the first-line treatment for spinal compression until the cause of the compression is determined. Dexamethasone may be administered at a dose of 2 mg/kg every six hours or 30 mg/kg for the first eight hours. Corticosteroids reduce oedema by suppressing inflammation and stabilising vascular membranes in the medulla/spinal cord, thereby alleviating pain and other neurological symptoms.[9] Our patient was administered 30 mg/kg Methylprednisolone as the final diagnostic intervention.

Emergency treatment is required for patients with rapidly progressing neurological symptoms. In cases of rapid onset, compression should be corrected within 8–10 hours, whereas for compression that develops over a 24–28-hour period or longer, decompression within seven days is sufficient. The treatment choice depends on the type of tumour causing the compression.[8, 12]

Given the rapid progression of paraplegia in our patient, we opted to perform surgery.

Conclusion

We diagnosed and treated a rare case of ES in which motor and sensory deficits on neurological examination were suggestive of GBS; however, imaging and pathology findings revealed the correct diagnosis of ES. It shows that spinal cord compression can be diagnosed and treated rapidly in the emergency department. Corticosteroid therapy should be initiated until a treatment plan is developed.

Disclaimer: This work was presented as an oral presentation at the 6th Intercontinental Emergency Medicine Congress and 6th International Critical Care and Emergency Medicine Congress that was held in Antalya, Turkey, April 25-28, 2019.

Conflict of interest: None to declare.

Funding disclosure: None to declare.

References

1. Willison HJ, Jacobs BC, van Doom PA. Guillain-Barré syndrome. Lancet 2016; 388 : 717:7.

2. Van Doorn PA. Ruts L. Jacobs BC. Clinical features, pathogenesis, and treatment of Guillain Barré syndrome. Lancet Neurol 2008; 7: 939-50.

3. Gajjar MD, Bhatnagar NM, Patel NJ, Patel T. Guillain - Barre syndrome in a patient with acute myocardial infarction with ventricular septal defect repair treated with plasma exchange. Asian J Transfus Sci 2015; 9: 87-8.

4. Bernstein M, Kovar H, Paulussen M, Randall RL, Schuck A, Teot LA, et al. Ewing's sarcoma family of tumors: current management. Oncologist 2006; 11: 503-19.

5. De Ioris MA, Prete A, Cozza R, Podda M, Manzitti C, Pession A, et al. Ewing sarcoma of the bone in children under 6 years of age. PLoS One 2013; 8: e53223.

6. Kissane JM, Askin FB, Foulkes M, Stratton LB, Shirley SF. Ewing's sarcoma of bone: clinicopathologic aspects of 303 cases from the Intergroup Ewing's Sarcoma Study. Hum Pathol 1983;14: 773-9.

7. Lewis DW, Packer RJ, Raney B, Rak IW, Belasco J, Lange B. Incidence, presentation, and outcome of spinal cord disease in children with systemic cancer. Pediatrics 1986; 78: 438-43.

8. Prasad D, Schiff D. Malignant spinal-cord compression. Lancet Oncol 2005; 6: 15–24.

9. Rheingold SR, Lange BJ. Oncologic Emergencies. In: Pizzo PA, Poplack DG. Principal and Practice of Pediatric Oncology. Philadelphia: Lippincott Williams & Wilkins. 2006; 1202-30.

10. Kelley KM, Lange B. Oncologic emergencies. Pediatr Clin North Am 1997; 44: 809-30.

11. Hayes FA, Thompson EI, Hvizdala E, O'Connor D, Green AA. Chemotherapy as an alternative to laminectomy and radiation in the management of epidural tumor. J Pediatr 1984; 104: 221-4.

12. Pashankar FD, Steinbok P, Blair G, Pritchard S. Successful chemotherapeutic decompression of primary endodermal sinus tumor presenting with severe spinal cord compression. J Pediatr Hematol Oncol 2001; 23: 170-3.

Figure-1: Brain computed tomography showing the hyperdense area adjacent to the right frontal lobe (arrows).

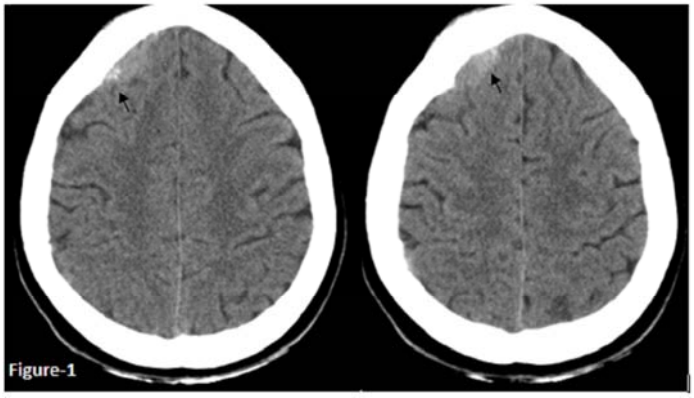
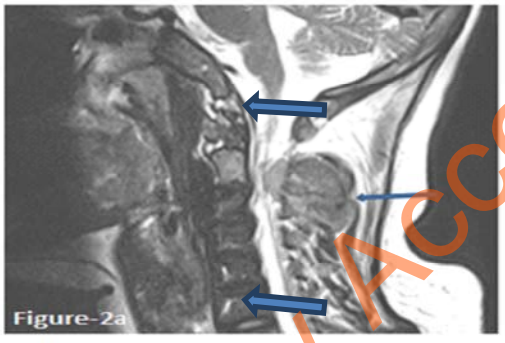
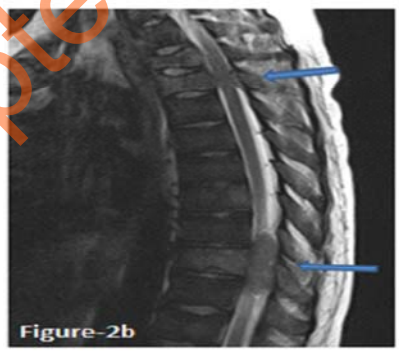
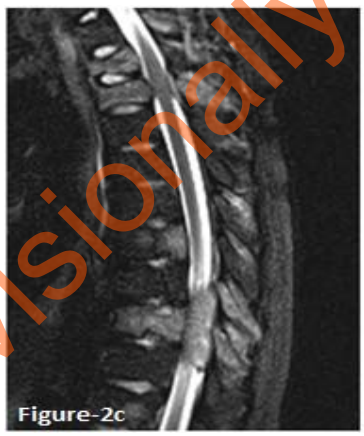


Figure-2: Spinal magnetic resonance images showing multiple mass lesions surrounding the spinal vertebrae at C2–3 (a) and T1–2 and T6–8 (b). Contrast-enhanced images of the lesions are shown at T1–2 and T6–8 (c), along with comparison of the lesions at T6–8 in the traverse and longitudinal planes (d).







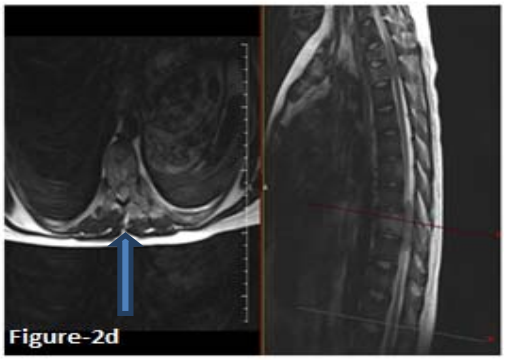
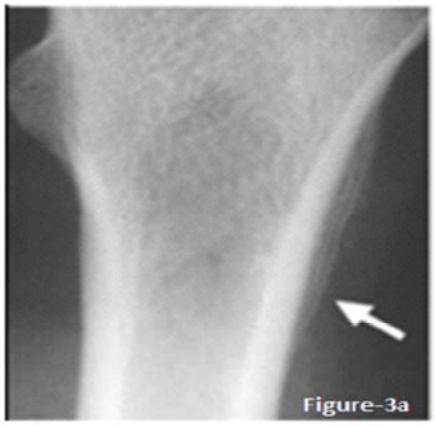
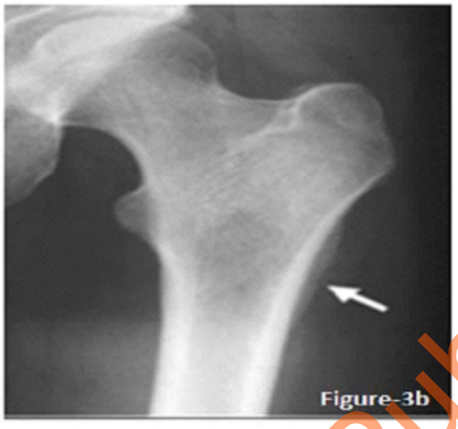


Figure-3: Direct radiographs showing lytic lesions in the proximal left femur (a) and pelvis (b).





The English in this document has been checked by at least two professional editors, both native speakers of English. For a certificate, please see:
http://www.textcheck.com/certificate/x8Jpo8

Patient consent form

For a patient's consent to publication of information about them in the *Journal of Pakistan Medical Association*

Title of article: An unusual presentation of Ewing sarcoma mimicking Guillain–Barré syndrome in a 17-year-old male

Corresponding author: Ozgur Sogut

I Huseyin Giray [insert full name] give my consent for this information about **MYSELF**/MY CHILD OR WARD/MY RELATIVE [circle correct description] relating to the subject matter above ("the Information") to appear in the Journal of Pakistan Medical Association (JPMA).

I have seen and read the material to be submitted to the JPMA

I understand the following:

(1) The Information will be published without my name attached and the JPMA will make every attempt to ensure my anonymity. I understand, however, that complete anonymity cannot be guaranteed. It is possible that somebody somewhere - perhaps, for example, somebody who looked after me if I was in hospital or a relative - may identify me.

(2) The text of the article will be edited for style, grammar, consistency, and length

(3) The Information may be published in the monthly JPMA, which is distributed worldwide. The journal goes mainly to doctors but is seen by many non-doctors, including journalists.

(4) The Information will also be placed on the JPMA's website,

(5) *The information may also be used in full or in part in other publications and products published by the JPMA or by other publishers to whom the JPMA licenses its content. This includes publication in English and in translation, in print, in electronic formats, and in any other formats that may be used by the JPMA or its licensees now and in the future.

(6) The JPMA will not allow the information to be used for advertising or packaging or to be used out of context (for example, a photograph will not be used to illustrate an article that is unrelated to the subject of the photograph.)

(7) I can revoke my consent at any time before publication, but once the information has been committed to publication ("gone to press") it will not be possible to revoke the consent.

Signed:

Date: 12.03.2020

227