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CASE REPORT

Retroperitoneal Ewing Sarcoma among Asian Patients: A Case Report and Review of Literature

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ABSTRACT

Retroperitoneal Extraosseous Ewing Sarcoma (EES) is a rare malignancy and poses diagnostic and management challenges. We present a case report of a 32-year-old Malay male who experienced persistent abdominal distension with constitutional symptoms. Imaging studies revealed an aggressive retroperitoneal mass with lymphadenopathy and local invasion. The diagnosis of retroperitoneal EES was confirmed through histopathological examination. The patient's condition persisted, but he refused surgical intervention. To address the persistent pain, the patient underwent coeliac plexus block and neurolysis. We also reviewed case reports regarding retroperitoneal EES for better insight into the management of this rare sarcoma.

KEYWORDS: Ewing Sarcoma; Rare sarcoma; Retroperitoneal Ewing Sarcoma; Coeliac Plexus Block; Asian Patients

INTRODUCTION

Retroperitoneal sarcoma is considered a rare cancer and an uncommon tumour. Among the sarcoma, Extraosseous Ewing Sarcoma (EES) in the retroperitoneal is categorized as an ultra-rare sarcoma [1]. It poses a diagnostic challenge due to its rarity and nonspecific symptomatology. In this case, the patient had multiple visits to the general practitioners and was treated as acute dyspepsia, until he decided to come to the emergency department. The incidental finding of the abdominal mass led to further evaluation and treatment. Despite undergoing chemotherapy and radiotherapy, the patient's condition persisted. The patient's refusal of surgical intervention and disease progression led to severe pain, necessitating the implementation of multimodal pain management strategies, including coeliac plexus block and neurolysis. We describe the challenges in managing this case and a review of case reports among Asian patients.

CASE PRESENTATION

A 32-year-old male presented with a four-month history of abdominal distension, which gradually worsened. He also experienced abdominal pain, described as dull in nature, associated with constitutional symptoms. He had a significant weight loss of 15 kg within three months. Despite multiple visits to general practitioners, he sought further evaluation by coming to the Emergency Department in January 2021. An incidental finding of an abdominal mass led to further evaluation. Ultrasound showed a massive intraabdominal mass with

a notable mass effect. A computed tomography (CT) scan revealed a large, heterogeneously enhancing left retroperitoneal solid mass that extended from the T7/T8 vertebral level to the L3/L4 level. The mass crossed the midline to the right side, exerting pressure on the abdominal aorta and inferior vena cava and displacing them toward the right side. Magnetic resonance imaging (MRI) of the abdomen showed a large left retroperitoneal mass with a necrotic centre that displaced and compressed the surrounding abdominal organs (Figure 1). Histopathological examination (HPE) of the left retroperitoneal mass biopsy, conducted by the surgical team, revealed monomorphic small blue round cells with distinct characteristics. These cells displayed round to oval hyperchromatic to vesicular nuclei, fine chromatin, and small inconspicuous nuclei with scanty to clear cytoplasm. The presence of glycogen was highlighted by periodic acid-Schiff (PAS) and PAS-Diastase (PASD) stains. Immunohistochemistry further supported the diagnosis, demonstrating strong diffuse membranous CD99 positivity, nuclear FLI-1 positivity, S100 positivity, and focal EMA staining. Collectively, these findings strongly support the diagnosis of ES.

Despite undergoing chemotherapy and radiotherapy, the patient's condition persisted. Additionally, despite explanations from the primary team, the patient declined any surgical intervention, expressing his fear of the intervention. To manage persistent pain, the patient underwent coeliac plexus block and neurolysis administered by the acute pain service (APS) team. Effective pain relief was achieved for six months until the pain recurred and could not be managed with opioid therapy. Follow-up imaging post-treatment showed that there was a slight reduction in the mass size. Unfortunately, there was a new enhancing pelvic mass indicating progressive disease (Figure 1).

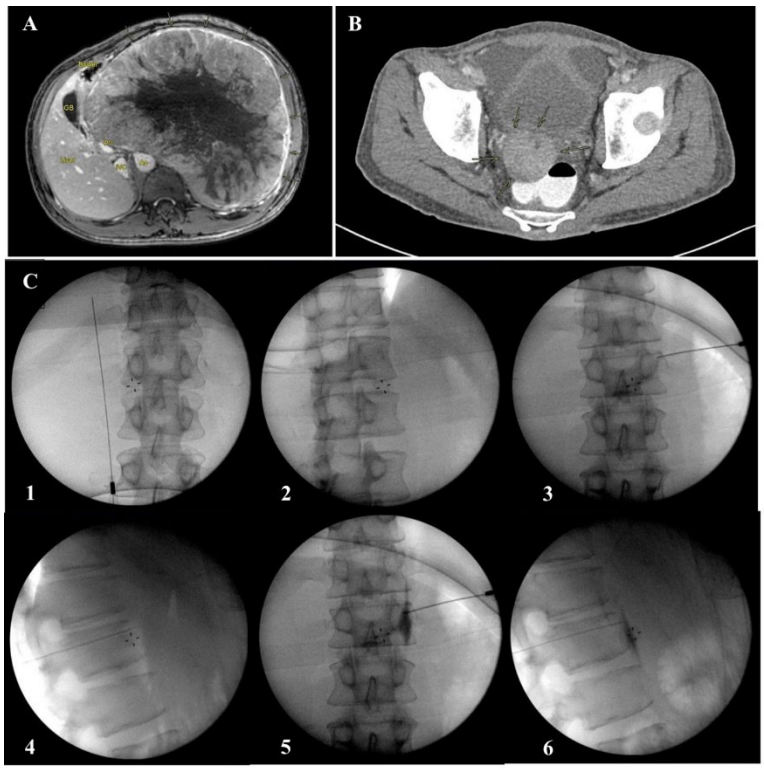

Figure 1 A) MRI of the abdomen showed a huge left retroperitoneal mass (yellow arrows) occupying most of the space within the abdomen. The mass displaces and compresses the aorta (Ao), inferior vena cava (IVC), portal vein (PV), liver, gallbladder (GB), and bowel, as seen in this figure. B) Contrast-enhanced computed tomography (CT) of the pelvis post-treatment showed a new enhancing mass anterior to the rectum (yellow arrows). This mass was not previously seen and indicates disease progression. C) Real-time images obtained during the CPB procedure by using C-arm Image Intensifier (II) 1) 12th rib, L1 vertebral body and transverse process identified (AP view) 2) 12th rib, L1 vertebral body and transverse process identified (lateral view) 3) Needle introduced by using right-sided posterior approach along the L1 vertebral body 4) Tip of needle bypasses the anterior border of L1 vertebral body 5) Contrast material used to visualize abdominal aorta border (AP view) 6) Contrast material used to visualize abdominal aorta border (lateral view)

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To provide persistent pain relief, a different approach was employed. A single-needle right-sided posterior coeliac plexus block (CPB) and neurolysis were performed under real-time C-arm Image Intensifier guidance. The procedure involved identifying the 12th rib, L1 vertebral body and L1 transverse process (Figure 1). The aorta is visualized with contrast material after the needle tip bypasses the anterior border of the vertebral body of L1 (Figure 1). It is followed by the introduction of a steroid solution comprising lignocaine 1% and triamcinolone acetonide 50 mg into the retro-aortic space. Neurolysis was then performed by injecting 10 ml of 70% alcohol into the same area. In our case, we utilized a single needle entry from the right approach only, as the mass had caused the aorta to shift to the right, occupying the left side. This approach was taken to avoid the risk of bleeding or hematoma should we have punctured through the mass on the left side. The patient underwent a pain-free interval, registering a numerical rating scale (NRS) pain score of 6 pre-procedure and achieving a score of 0 post-procedure, with no observed complications.

DISCUSSION

Ewing sarcoma (ES) is an aggressive form of bone cancer that primarily affects adolescents and young people. The incidence is higher in the Caucasian population, with only a few cases reported in the Asian community, and 80% of cases occur in individuals under the age of 18 [2]. It may also develop in various sites, such as extraosseous Ewing sarcoma (EES) in the retroperitoneum, which is categorized as an ultra-rare sarcoma [1]. Retroperitoneal EES symptoms tend to be non-specific and eventually grow to substantial sizes before detection [1].

The standard treatment approach for EES typically involves a multimodal approach, incorporating chemotherapy, surgery, and radiation therapy, tailored to the specific characteristics and location of the tumour.

The adoption of initial treatment with vincristine,doxorubicin, cyclophosphamide/ifosfamide, and etoposide (VDC/IE) followed by multi-visceral resection has emerged as the new standard of care [1]. Since this patient had severe pain, CPB was applied. CPB is a minimally invasive procedure designed for chronic upper abdominal pain management in patients with pancreatic cancer or related malignancies [3]. For individuals with retroperitoneal EES experiencing persistent pain despite pharmacological analgesic therapy, CPB is a valuable and safe treatment option. The procedure offers effective pain relief with minimal side effects and improve the quality for this specific patient cohort.

We identified 10 published case reports regarding retroperitoneal EES cases in Asian countries, summarized in Table 1. These cases primarily involved female patients, predominantly in the young adult age group. Surgery was the mainstay of treatment, with most patients experiencing successful outcomes and survival. There was no tumor recurrence and/or resolution during follow-up post-treatment in four of the cases [4-7]. One case reported the patient's demise due to disease progression [8]. The presentation of retroperitoneal EES varies, with many patients experiencing abdominal pain and/or a palpable mass during physical examination [6,8-10]. In one case, the tumour was incidentally discovered by MRI, despite normal physical examination findings [4]. The diagnosis mainly relies on histopathology and immunohistochemistry, showing small, rounded cells with positive CD99 and vimentin markers. Molecular and genetic profiling through fluorescence in situ hybridization (FISH) is crucial for accurate diagnosis, as Ewing sarcoma is characterized by rearrangement of the EWSR1 gene within tumour cell nuclei [1].

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Table 1 Summary of case reports

Case Report	Age/ Gender	Symptoms/Signs	Histopathology/ Immunochemistry	Intervention	Outcome
Shu-Yu Wu (2023) [4] (Taiwan)	57-year-old woman	Large retroperitoneal tumour detected by MRI, no visible or tender mass during physical exam.	Round blue cell tumour.	- Laparotomy with tumour resection - Radiation therapy - Chemotherapy	No tumour recurrence six months post-operation.
S.Chen (2022) [5] (China)	17-year-old male	Abdominal distension and discomfort for two months. Palpable mass in upper abdomen.	Postoperative pathological diagnosis is EES.	- Radical resection of tumour. - Chemotherapy - Radiotherapy	Liver metastasis was found by CT and partial response (PR) to treatment.
Wang JL (2022) [11] (China)	A 20-year-old female	Intermittent pain under the right costal arch for four months.	Tumour cells with round nuclei, pale cytoplasm, and rosette structures. Immunohistochemistry: positive for CD99 and Nkx2.2. Fluorescence in situ hybridization (FISH) studies confirm rearrangement of the EWSR1 gene in tumour cell nuclei.	- Tumour resection - Combination chemotherapy	Survived for 20 months since discharge with ongoing follow up.
R. AlRashed (2022) [9] (Saudi Arabia)	26-year-old woman	Progressive left-sided abdominal pain for four months, radiating to back and left loin, accompanied by fatigue. Palpable round, firm mass in left upper and lower quadrants.	Extensive necrosis, fibrosis, and haemorrhage. Focal residual round blue cell tumour. EWSR1 rearrangement positive by FISH.	- Neoadjuvant chemotherapy. - Laparotomy with tumour resection.	Post-operative recovery and discharged home on day five.
F.N. Nedham (2022) [8] (Bahrain)	35 years old male	Epigastric pain with melena. Early satiety and generalized fatiguability. Unintentional weight loss of approximately 10 kg. Epigastric tenderness without guarding or rigidity, no organomegaly or abdominal mass.	Diffusely arranged small round blue cells with minimal vascularized collagenous stroma. Tumour cells strongly positive for Vimentin and CD99, moderately positive for BCL2, weakly positive for CD117. Ki67 shows a 50% proliferation index.	-Gastro-jejunostomy bypass to alleviate compression of the duodenum caused by the tumour. -Chemotherapy.	Passed away 15 months from initial presentation.
Anita P Javalgi (2016) [10] (India)	39-year-old female	Abdominal discomfort and vague pain since two months, accompanied by weakness in the lower limb and weight loss.	Small round blue cell tumor with PAS positivity. Tumour shows positive staining for S100, Vimentin, and CD99.	- Laparotomy. - Chemotherapy.	Lost to follow-up.
S. Ulusan (2007) [12] (Turkey)	26-year-old woman	Swelling in the left inferior abdomen for one month duration. Palpable abdominal mass on the left inferior side.	Tumour cells exhibited a small, solid, lobular pattern with uniformity. The cytoplasm was pale and scanty. Immunohistochemically, Vimentin showed patchy	- Surgical removal. - Adjuvant chemotherapy.	NA

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			positivity, and CD99 stained positive.		
SK Chang (1995) [13] (Singapore)	27-year-old Chinese male	Syncopal attack. Cough and right flank discomfort for several days.	Soft, friable tumour with necrotic areas. Microscopically, small round cells with indistinct cytoplasm and distinct nuclei. Occasional rosette-like structures. Strong vimentin staining.	- Laparotomy with tumour removal. - Chemotherapy.	Patient recovered well post-surgery. Significant resolution of the residual retroperitoneal tumour mass.
M. Oya (1995) [6] (Japan)	34-year-old Male	Left flank pain. Left costovertebral angle tenderness on physical examination.	The tumour consisted of closely packed small round cells with indistinct cell borders. Contained fine cytoplasmic granules that stained positive with the PAS reaction. Immunohistochemistry demonstrated positive reactivity for Vimentin and 5C11.	- Radical extirpation of tumour. - Adjuvant chemotherapy.	No recurrence or other symptoms at 10 months follow up.
Hiroshi Hara (1991) [7] (Japan)	18-year-old Female	Hematuria detected during an examination two years ago when the patient complained of right lumbar pain. The patient presented to hospital in June 1988 with a chief complaint of a right abdominal tumour.	Histopathological diagnosed as extra skeletal ES.	-Surgically removed	No recurrence after two years with on-going follow up.

CONCLUSION

Clinical findings of abdominal mass, aided by imaging and prompt treatment, may significantly decrease morbidity and mortality in retroperitoneal EES patients. While CPB may not influence the prognosis of EES, it is an alternative approach for managing severe and persistent pain in affected patients.

Conflict of Interest

Authors declare none.

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Author Contributions

(1) Concept or design: All authors
(2) Acquisition of data: All authors
(3) Analysis or interpretation of data: All authors
(4) Drafting of the article: All authors
(5) Critical revision for important intellectual content: All authors
(6) Final approval of the version published: All authors

All authors had full access to the data, contributed to the study, approved the final version for publication, and take responsibility for its accuracy and integrity.

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