Case Report

Clear Cell Sarcoma of the Pelvis in a Male Child, Diagnosed Using Ancillary Techniques - A Rare Case Report

Pushpit Gupta[1], Bembem Khuraijam[1,*], Varuna Mallya[1], Shaad Vali[1], Sujoy Neogi[2]

[1]Department of Pathology, Maulana Azad Medical College, New Delhi, India

[2]Department of Paediatric Surgery, Maulana Azad Medical College, New Delhi, India

***Correspondence:** bembemkh@yahoo.com

DOI
10.21276/apalm.3675

Article History
Received: 15-08-2025
Revised: 23-11-2025
Accepted: 17-12-2025
Published: 05-01-2026

How to cite this article
Gupta P, Khuraijam B, et al. Clear Cell Sarcoma of the Pelvis in a Male Child, Diagnosed Using Ancillary Techniques - A Rare Case Report. Ann Pathol Lab Med. 2026;13(1):C8-C13.

Copyright

This work is licensed under the Creative Commons Attribution 4.0 License. Published by Pacific Group of e-Journals (PaGe).

Abstract

Clear cell sarcoma is a rare malignant neoplasm of uncertain differentiation that typically arises in the deep soft tissues, most frequently involving the tendons and aponeuroses of the extremities in young adults, particularly females. Diagnostic challenges often arise due to its striking histological and immunohistochemical resemblance to metastatic malignant melanoma. We report an exceptionally rare case of clear cell sarcoma occurring in a nine-year-old male child, located in the pelvis with direct extension into the right pelvic bone. Non-Contrast Computed Tomography (NCCT) and Magnetic Resonance Imaging (MRI) demonstrated tumour extension into the presacral space with intraspinal involvement, resulting in widening of the spinal canal and neural foramina. Trucut biopsy revealed nests of epithelioid to spindle-shaped cells with clear to eosinophilic cytoplasm and prominent nucleoli. Bone marrow aspiration from the right iliac region yielded tumour cells, confirming marrow infiltration. The diagnosis was established through a multimodal approach integrating radiological assessment, bone marrow aspiration cytology, and trucut core biopsy findings. Ancillary studies, including flow cytometry, immunohistochemistry, and special stains, further supported the diagnosis in this challenging presentation. A review of the relevant literature is provided, highlighting distinguishing features from morphologically similar lesions to enhance recognition of this aggressive, metastasis-prone malignancy.

Keywords: clear cell sarcoma; pelvis; male; child

Introduction

Clear cell sarcoma (CCS) is a malignant soft tissue tumour currently classified under “tumours of uncertain differentiation” in the fifth edition of the World Health Organization (WHO) Classification of Soft Tissue and Bone Tumours.[1] It predominantly arises in the deep soft tissues, with a particular predilection for the tendons and aponeuroses of the extremities, and only rarely occurs in the head, neck, or trunk regions.[2] Owing to its significant histopathological and immunohistochemical resemblance to malignant melanoma, CCS poses considerable diagnostic challenges and permits only a narrow margin for diagnostic error.

CCS typically presents at a mean age of approximately 30 years and shows a modest female predominance. Its overlapping histomorphological features with other soft tissue neoplasms—including synovial sarcoma, malignant peripheral nerve sheath tumour, perivascular epithelioid cell neoplasm, epithelioid sarcoma, and paraganglioma-like dermal melanocytic tumour—often necessitate the demonstration of EWSR1::ATF1 fusion or EWSR1 gene rearrangement for definitive diagnosis.[3]

Here, we present a case characterized by an atypical clinical presentation, in which the final diagnosis was established only after comprehensive evaluation using multiple diagnostic modalities.

Case Report

A nine-year-old male child presented with complaints of back pain and right lower-limb pain for the past five months, along with right lower-limb disuse atrophy, presacral swelling, and a limping gait for the past two months. On examination, a palpable lump was noted in the sacral region. It was ill-defined, firm in consistency, and non-mobile. The overlying skin showed no ulceration or other changes. Systemic examination revealed no pallor, icterus, or lymphadenopathy. Cardiovascular and respiratory system examinations were unremarkable. Neurological examination showed mildly reduced power (four/five) in the right lower limb.

After examination, Non-Contrast Computed Tomography (NCCT) was done, which revealed an ill-defined, destructive soft tissue mass involving the sacrum, extending into the presacral space predominantly on the right side, with intraspinal extension causing widening of the spinal canal and neural foramina. Also, Magnetic Resonance Imaging (MRI) of the pelvis demonstrated a large, uniformly enhancing solid soft tissue mass centred in the presacral space, with associated involvement of the sacrum, widening of the sacral neural foramina, and extension into the spinal canal with indistinct fat planes. Based on the NCCT and MRI findings—specifically the extension through the neural foramina producing a dumbbell-shaped lesion, ill-defined osteolytic destruction, widened foramina, and spinal canal involvement—a provisional suspicion of neuroblastoma or an Ewing sarcoma family tumour was raised.

Based on the suspicion of Neuroblastoma/Ewings family of tumours, biochemical investigations, followed by a trucut biopsy of the mass, including immunohistochemical staining was done. At the same time, bone marrow aspiration studies, including bone marrow aspirate in EDTA (Ethylenediaminetetraacetic acid) and flow cytometry, were performed to check for involvement, as the lesion appeared to be invading the right iliac bone. Serum Lactate Dehydrogenase (LDH) levels were elevated at 261 U/L (Reference range: 125-220 IU/L), while Serum α-Fetoprotein (AFP) levels were within normal limits at 1.69 ng/mL (Reference range: $<$10 ng/mL).

The trucut biopsy revealed nests of epithelioid to spindle-shaped cells with clear to eosinophilic cytoplasm and prominent nucleoli. The tumour cells were arranged within a delicate fibro-collagenous stroma, some areas of which contained blood vessels. The individual cells were oval to elongated, cytologically monotonous, and exhibited occasional mitoses. Focally scattered melanin pigment was noted within some tumour cells. This pigment was confirmed as melanin by positive staining with the Masson–Fontana technique. The tumour cells showed immunopositivity for Vimentin, S100, and faint positivity for HMB-45, while they were negative for CK, Desmin, MyoD1, Synaptophysin, NSE, CD99, and LCA.

In view of the absence of small round blue cell morphology, the presence of melanin pigment, and the lack of immunoreactivity for Synaptophysin, NSE, and CD99, a diagnosis of neuroblastoma or an Ewing sarcoma family tumour was considered unlikely. Figure 1 illustrates the trucut biopsy features and NCCT findings.

Bone marrow aspiration cytology from the left side demonstrated particulate, normocellular marrow with normal maturation of the erythroid, myeloid, and megakaryocytic lineages. In contrast, the right-sided aspirate yielded no marrow particles but revealed tumour deposits composed of sheets and singly scattered atypical cells with a high nucleocytoplasmic ratio, scant to moderate pale basophilic cytoplasm, and irregular nuclear contours. Some cells displayed prominent nucleoli, and a few binucleate forms and pigment-containing cells were also identified.

The right-sided bone marrow aspirate in EDTA was subjected to flow cytometric analysis. Flow cytometry was performed on a 10-color/3-laser Navios EX-Beckman Coulter flow cytometer, and the analysis was performed using Kaluza software. The instrument calibration, quality control, voltage and compensation were performed as per the manufacturer's instructions. After removing doublets and debris, viable cells were separated which were gated using CD45 vs Side Scatter strategy and CD45 negative cells were gated.

Flow cytometry revealed a distinct population of CD45-negative cells exhibiting moderately strong CD56 expression. An unstained tube was put alongside to check for false positive expression. Bone marrow trephine biopsy was not performed in this case due to patient discomfort and expected diagnostic sufficiency from bone marrow aspiration. Figure 2 presents the flow cytometry and cytology findings of this case.

On further workup, no additional lesions were identified in the skin, sun-exposed areas, iris, gastrointestinal tract, anogenital

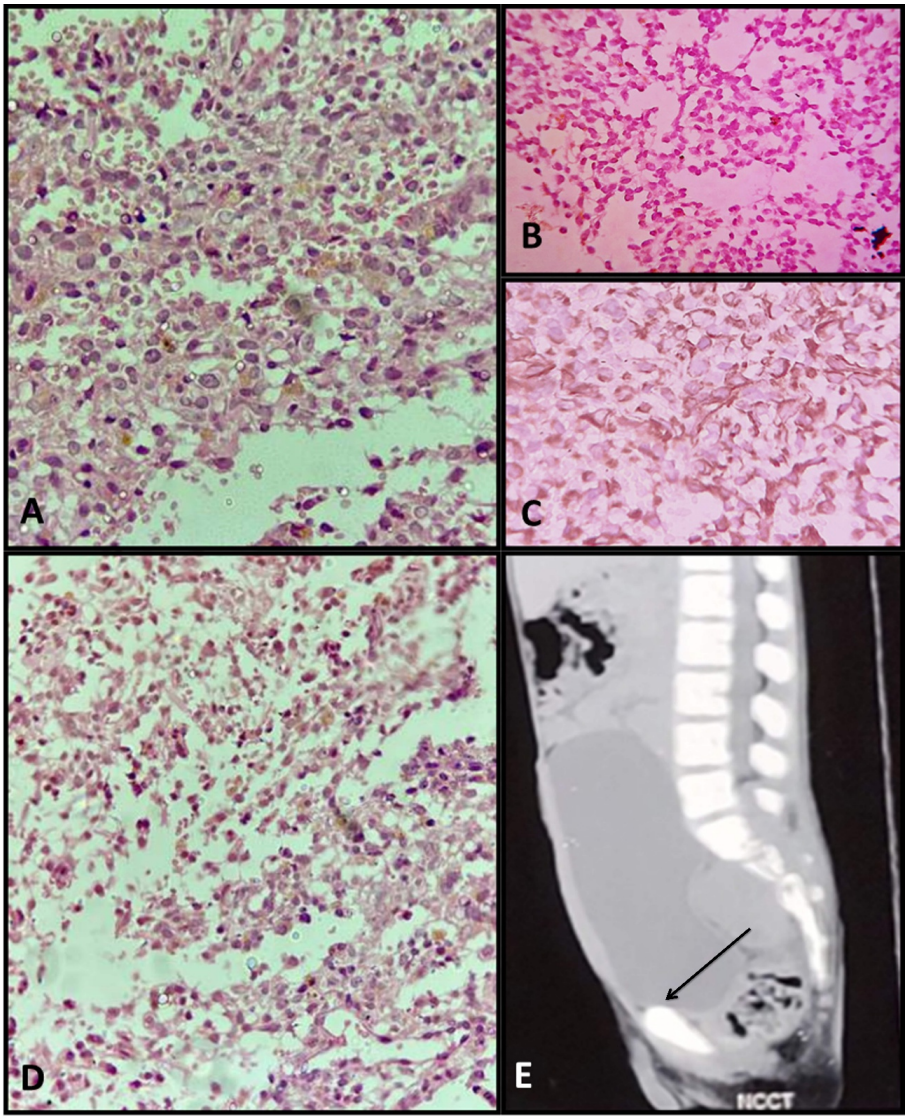

Figure 1: A, D- Trucut biopsy shows sheets of tumour cells with round to oval epithelioid morphology, eosinophilic to clear cytoplasm, occasional prominent nucleoli, and scattered melanin pigment (H&E, 400×). B- Immunohistochemistry for HMB-45 showing faint positivity (IHC, 400×). C- Immunohistochemistry for Vimentin showing strong positivity (IHC, 400×). E- Coronal section of NCCT showing the tumour in the pelvis with invasion through the right pelvic bone (arrow).

region, sinonasal tract, genitourinary tract, or leptomeninges.

Discussion

The case presented here is exceedingly rare due to its occurrence in a male child under ten years of age and its presentation at an uncommon pelvic site. For literature search, PubMed / PMC (via NCBI), Google Scholar, ResearchGate, Publisher pages / journal websites and broad web search to capture conference reports/small journals not in PubMed were done. For PubMed search, the following code was used - (”Clear Cell Sarcoma”[MeSH] OR ”clear cell sarcoma” OR ”malignant melanoma of soft parts”) AND (child OR pediatric OR paediatric OR ”child*”[MeSH]) AND (male) AND (pelvis OR pelvic OR sacrum OR iliac OR presacral). Similarly, other databases were also searched. To the best of our knowledge, based on a systematic search of PubMed, PMC, Google Scholar, and publisher websites till date, this appears to be the first reported case of clear cell sarcoma involving the pelvis/pelvic bone in a male child under ten years of age in the English-language literature.

A noteworthy finding in this case was the direct involvement of the right iliac bone by the tumour, which resulted in tumour deposits being obtained on bone marrow aspiration. Bone marrow aspiration and flow cytometry were performed due to the clinical suspicion of Neuroblastoma/Ewings family of tumours and flow cytometry aided by excluding a haematolymphoid malignancy involving bone marrow, further characterizing immunophenotype (CD45 negative, CD56 positive) in conjunction with immunohistochemistry on trucut biopsy and assessing the extent of bone marrow involvement by tumour.

CD56 expression has been reported in approximately 21% of clear cell sarcoma cases in the literature;[4] however, these data are derived from immunohistochemical analysis of tissue samples. There is no established flow cytometric immunophenotypic

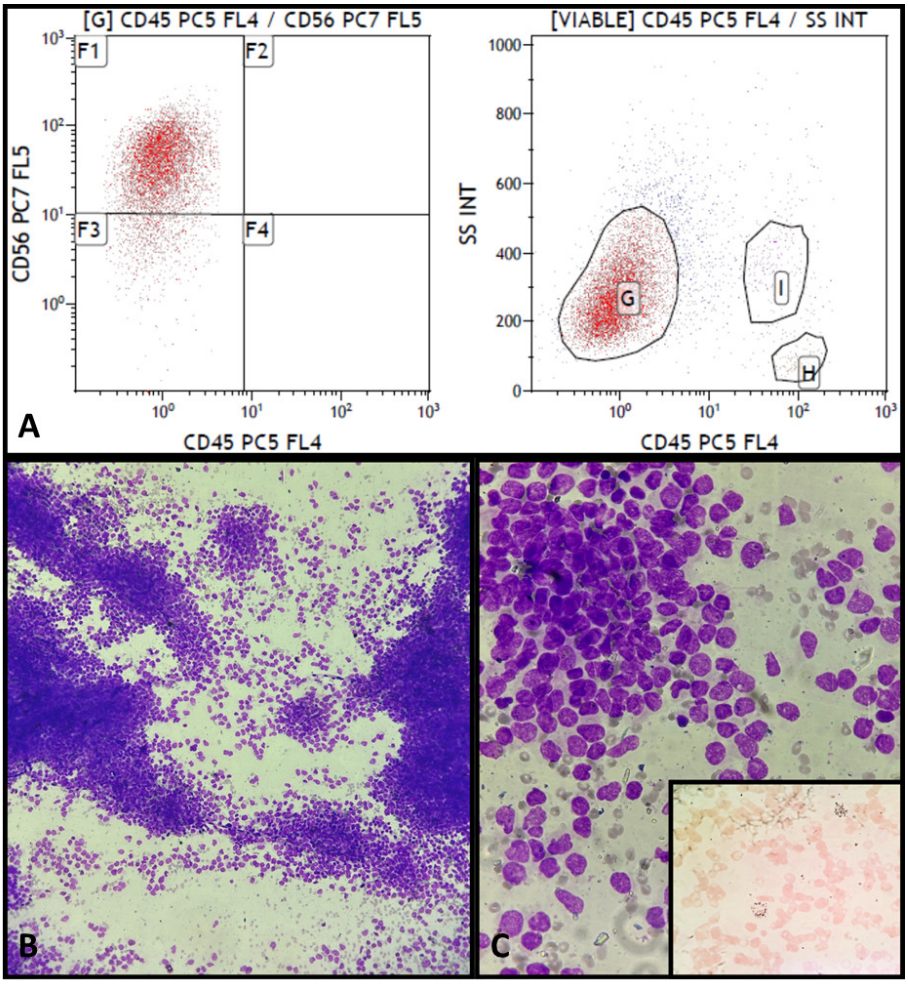

Figure 2: A - Right image showing gating of CD45-negative tumour cells using a CD45 vs. side-scatter gating strategy, while the left image shows weak to moderate CD56 expression in the tumour cells. B - Tumour cells arranged in large clusters as well as scattered singly in the bone marrow aspirate from the right side (Giemsa, 100×). C - Individual tumour cells showing round to oval epithelioid morphology with irregular nuclear contours and a high N:C ratio (Giemsa, 400×). Inset shows positivity for Masson Fontana special stain in tumour cells (400x).

profile that reliably differentiates clear cell sarcoma (CCS) from malignant melanoma (MM). Nonetheless, a study by el-Naggar et al. explored the use of DNA content analysis by flow cytometry in attempting to distinguish CCS from MM.[5]

However, demonstration of the EWSR1::ATF1 fusion or EWSR1 gene rearrangement is required for a definitive diagnosis, due to unavailability of specific Polymerase Chain Reaction (PCR) primers/probes, Fluorescent in-situ Hybridization (FISH) or other molecular assays, the test could not be performed in this case. Hence, the diagnosis was established through an integrated approach combining clinical features, radiological imaging, cytology, histopathology, immunohistochemistry, and flow cytometry.

A previous case report by Nakayama et al. documented clear cell sarcoma of the pubic bone in an 81-year-old man, in which the tumour exhibited morphology resembling a small round cell neoplasm, highlighting the diagnostic challenges posed by its variable histological appearance.[6] Table 1 provides a comparison of the differential diagnoses considered for this tumour in relation to the present case.

Another report by Wasnik et al. described a clear cell sarcoma arising in the pelvis of a 26-year-old man who presented with lower-limb weakness, muscle wasting, foot drop, and an infiltrative mass on imaging.[7]

Conclusion

Although clear cell sarcoma (CCS) typically presents within a characteristic age group and at well-defined anatomical sites, a high index of suspicion should be maintained when evaluating lesions at atypical locations. Particular emphasis must be placed on distinguishing CCS from other morphologically similar entities, especially metastatic malignant melanoma. A thorough evaluation to exclude primary melanoma—including assessment of the skin, iris, and other melanoma-prone sites—is essential. Optimal management with long-term surveillance for distant metastasis is required, as many patients experience disease-related mortality due to metastatic spread.

Table 1: Various differential diagnoses of the tumour in comparison with the present case.

Differentials	Age Group	Location	Histopathological Characteristics	Immunohistochemistry
Clear Cell Sarcoma – Usual Case	Adults	Arms, legs, feet and hands most commonly.	Infiltrative cells in compact nests and fascicles that dissect along with dense connective tissue with clear to eosinophilic cytoplasm.	Positive for S-100, SOX10, HMB45. Negative for SMA, DESMIN.
Epithelioid Sarcoma	Adolescents and young adults	Anywhere. Distal extremities (classic type), truncal tissue (proximal type)	Classical type – simulate granulomatous process with or without necrosis. Proximal type – infiltrative growth with large polygonal cells.	Positive for Pan-CK, Vimentin, CA-125. Negative for S-100.
Perivascular Epithelioid Cell Neoplasm	Adults	Lungs, GI tract, kidneys, liver, uterus etc.	Perivascular epithelioid cells with clear to eosinophilic cytoplasm and round to oval nuclei in a nested or trabecular arrangement. Lesser nuclear atypia than CCS.	Positive for HMB 45, muscle markers and Negative for S-100.
Malignant Peripheral Nerve Sheath Tumor	Adults	Anywhere. Trunk and extremities are most common.	Uniform spindle cells with hyperchromatic, thin, wavy or focally buckled nuclei. Marbled appearance because of alternating hypo and hypercellular areas.	Positive for S-100, SOX10.
Synovial Sarcoma	Adults	Anywhere in body	Could be Biphasic (spindle cells and gland like epithelial structures) or Monophasic (hypercellular fascicular with little intervening stroma).	Positive for TLE1, CK7, CK19, BCL2, CD99.
Melanoma	Adults	Skin, uvea, Anorectal region, leptomeninges, sinonasal tract	Asymmetrical and poorly circumscribed lesions with architectural disturbance and usually marked cytological atypia.	Positive for S-100, HMB 45, MELAN A.

Consent for Publication: Written informed consent for publication of the clinical details and images was obtained from the patient's legal guardian, with assurance that the child's identity will remain confidential.

Acknowledgements: None

Funding: None

Competing Interests: None

References

1. Tahiri Elousrouti L, Hammas N, Elmernissi FZ, Elfatemi H, Chbani L. Clear-Cell Sarcoma With an Unusual Presentation Mimicking Metastatic Melanoma. Cureus. 2022 Nov 29;14(11):e32010.
2. Schöffski P, Wozniak A, Stacchiotti S, Rutkowski P, Blay JY, Lindner LH, et al. Activity and safety of crizotinib in patients with advanced clear-cell sarcoma with MET alterations: European Organization for Research and Treatment of Cancer phase II trial 90101 'CREATE'. Ann Oncol. 2017 Dec 1;28(12):3000-8.
3. Mavrogenis A, Bianchi G, Stavropoulos N, Papagelopoulos P, Ruggieri P. Clinicopathological features, diagnosis and treatment of clear cell sarcoma/melanoma of soft parts. Hippokratia. 2013 Oct;17(4):298-302.
4. Hisaoka M, Ishida T, Kuo TT, Matsuyama A, Imamura T, Nishida K, et al. Clear cell sarcoma of soft tissue: a clinicopathologic, immunohistochemical, and molecular analysis of 33 cases. Am J Surg Pathol. 2008 Mar;32(3):452-60.
5. el-Naggar AK, Ordóñez NG, Sara A, McLemore D, Batsakis JG. Clear cell sarcomas and metastatic soft tissue melanomas. A flow cytometric comparison and prognostic implications. Cancer. 1991 Apr 15;67(8):2173-9.
6. Nakayama S, Yokote T, Iwaki K, Akioka T, Miyoshi T, Hirata Y, et al. A rare case of primary clear cell sarcoma of the pubic bone resembling small round cell tumor: an unusual morphological variant. BMC Cancer. 2012 Nov 21;12:538.
7. Wasnik PD, Hussain N, Bodhey NK, Pati S. Clear cell sarcoma in pelvic region - case report. Indian Journal of Pathology and Oncology. 2021 May 15;8(1):162–5.