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Case report

Retroperitoneal blue cell round tumor (Ewing sarcoma in a 35 years old male)- case report

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ABSTRACT

Introduction: Ewing sarcoma is a highly aggressive tumor. It's common presentation is primary bone tumor, but very rarely as soft tissue sarcoma both of which are under the spectrum of Ewing sarcoma family of tumors (EFT).
Case presentation: A 35 years old male who presented with advanced locally spreading tumor. He was diagnosed after presenting with epigastric pain and melena, CT abdomen revealed the retroperitoneal mass. CT guided core biopsy taken and was diagnosed as Ewing sarcoma. The patient presented with mass already grown so he was not a surgical candidate. Patient was managed symptomatically till he was sent for palliative care in a Sarcoma centre in Jordon and passed away on October 2021.
Discussion: Ewing Sarcoma family of tumors are group of small round blue cell tumors that are histogenetically related. The better known is Ewing 's sarcoma (EWS). It is a malignant small blue round cell tumor with variable degree of neuroectodermal differentiation. Retroperitoneal tumor are mostly malignant and accounts for one third of soft tissue sarcoma. They usually present as large masses at the time of the diagnosis. So they do not produce symptoms until they grow large enough to compress or invade contagious structures.
Conclusion: ES-EWS is an aggressive tumor with high incidence of local recurrence and distant metastasis that's why is was given its poor prognosis characteristic. Multimodality treatment including surgical resection, chemotherapy and High dose radiotherapy will help in better survival rate.

1. Introduction

Ewing's Sarcoma is a rare type of highly aggressive tumor that usually presents as an undifferentiated primary bone tumor; less commonly presenting in soft tissue (the so called Extra-Skeletal/Extra- Osseous Ewing's Sarcoma). Both are part of a spectrum of neoplastic disease known as the Ewing sarcoma family of tumors(EFT). They represent a family of morphologically similar small round cell neoplasm that has an aggressive course and highly responding to combined multimodal treatment. The following reported case is an EWS in the retroperitoneal space in a young healthy male.

2. Case presentation

A 35 years old male not known of any chronic medical illnesses. Patient was initially admitted under internal medicine care in July 2020 due to Epigastric pain with melena. The pain was in the epigastric region for one month duration. It was not radiating. Associated with melena of around 4 times per day. He declared using ibuprofen and NSAIDs for his dental pain. No History of hematemesis, no recent use of any anti-coagulation or iron supplements, no History of dizziness, palpitation or loss of consciousness but he had History of early satiety and generalized fatiguability. He noticed non intentional weight reduction around 10 Kg in one month period. Those symptoms were associated with recurrent vomiting, which was bilious and after each oral intake. He is nonsmoker, Does not consume alcohol and is newly married and having one child. No significant Family or psychosocial history On Examination: patient was vitally stable, afebrile, looks conscious oriented, well hydrated not pale and not jaundiced. Abdominal examination revealed epigastric tenderness with on guarding or rigidity, no obvious organomegaly or

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abdominal mass. Investigation were done as follows: Hb was 10.9 mg/dl, Normal coagulation profile, Renal function, electrolytes and liver enzymes were within reference range. So the initial work up was to rule out upper or lower GI Bleeding. He Underwent Colonoscopy reach till the ileocecal junction and upper gastroscopy reaching down to the second part of the duodenum by gastroenterologist consultant on the same admission and were only significant for chronic active gastritis with no active bleeding seen and no masses seen, for which he was started on proton pump inhibitor BD intravenously. Given these endoscopic funding with the history of epigastric pain that did not settle, vomiting and weight loss CT abdomen with IV and Oral contrast done for him with some differentials in mind as duodenal lesion, Retroperitoneal lesion that is causing gastric outlet obstruction the CT scan showed the following: big retroperitoneal mass 10x9x8 cm encasing the aorta, compressing the IVC and duodenal third part (Image 1). The patient remained symptomatic with continuous vomiting so initial diagnosis of Gastric Outlet obstruction was made secondary to the retroperitoneal mass and is was referred to surgical team. Differentials were kept in mid as sarcoma, lymphoma, Pheochromocytoma. Pheochromocytoma workup were sent and came back -ve (Urine and serum catecholamines were normal. The next step was to diagnose the mass with tissue Biopsy, to know the degree of gastric outlet obstruction and to screen for any distant metastasis. So barium meal and follow through done to delineate the degree of obstruction and to plan for surgical diversion if needed. The fluoroscopic study showed partial obstruction at the level of the third part of the duodenum (Image 2).

After that tissue diagnosis was planned and he underwent CT Guided True Cut Biopsy on 12/8/2020 of the retroperitoneal mass by the interventional radiology consultant The procedure went smooth while he was admitted with uneventful recovery, remained stable hemodynamically and Subjectively. The microscopic Examination Showed mainly pancreatic tissue with few tiny fragments of tumor tissue which showed diffusely arranged small round blue cells with minimal vascularized collagenous stroma (Image 3). The tumor cells had minimal clear to eosinophilic cytoplasm and mild pleomorphism. The tumor cells were strongly positive for Vimentin and CD99, Moderately positive for BCL2 and Weakly positive for CD117. The Ki67 showed a 50% proliferation Index. The tumor cells were negative for CD3, CD20, synaptophysin, Chromogranin, CK7, CK20, S100, and Desmin - The overall Histopathological and Immunohistochemical appearance were consistent with Ewing Sarcoma. Metastatic screening including CT scan of the chest and liver were negative.

Patient case was discussed in the tumor board committee and he was planned to be managed in a sarcoma specialized centre. Meanwhile in 24th August 2020 (one month later after his medical admission) he was admitted to the hospital as he was not tolerating orally and developed prerenal azotaemia, recurrent drop in Hb and the need for Blood transfusions and Hydration. As the tumor was compressing Further on the Duodenum. Initial management were Hydration, NGT decompression and blood transfusion as mentioned then he underwent decompression by gastrojejunostomy bypass done by general surgery consultant on 26th August 2020 which was tolerated well Post-operative stay was in the ward basis with no acute events and was started on oral feeding after removal of the NGT on the second post-operative day. He was discharged then in a stable condition after tolerating feeds well and Improvement in his renal parameters. Afterwards he was doing fine till he was sent for further treatment in Jordon Sarcoma Centre. There he recieved around 6 cycles. His condition was deteriorating with no benefit. He passed away on 1st October 2021.

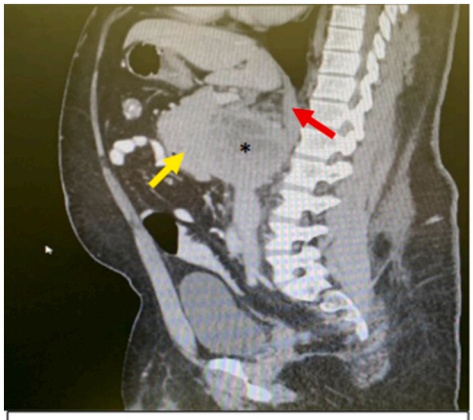

Image 1. CT abdomen showing retroperitoneal mass (*) with close proximity to the IVC posteriorly (red arrow) and the duodenum compression anteriorly (yellow arrow).

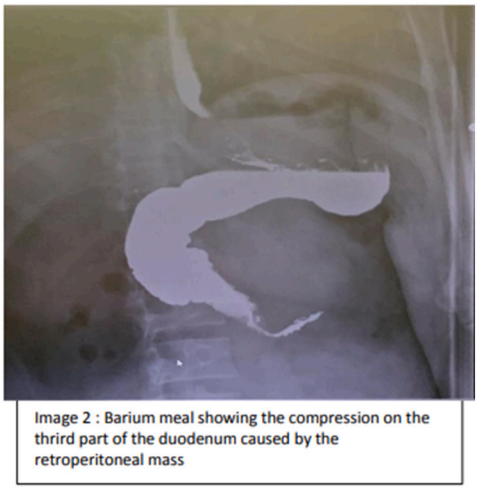

Image 2. Barium meal showing the compression on the third part of the duodenum caused by the retroperitoneal mass.

3. Discussion

Ewing Sarcoma family of tumors are group of small round blue cell tumors that are histogenetically related, all demonstrating nonrandom t (11:22)(q24:q12) chromosome rearrangement resulting in the formation of EWS-ETS Fusion Gene [1–3]. The better known of the Ewing sarcoma family is Ewing's sarcoma (EWS)/Peripheral NeuroEctodermal Tumor (PNET). It is a malignant small blue round cell tumor with variable degree of neuroectodermal differentiation [4,5]. Most common site of Ewing sarcoma is the bone and hence the so called skeletal EWS predominantly occurring in pediatric age group, while the extra- skeletal (extraosseous) EWS is very rare. In more than 50% of cases in adulthood Ewing's will be extra- skeletal. Sites that will grow the tumor would be variable such as trunk, intrabdominal tissues or retroperitoneum and the viscera accounting for 14%, 14% and 8%, respectively. So it can show us how rare is the extra-skeletal EWS [5,6]. They often grow rapidly with widespread metastasis by the time patient is presenting leading to the poor prognosis [7]. Retroperitoneal tumor are mostly malignant and accounts for one third of soft tissue sarcoma (10–15% of all adult tissue sarcoma). They usually present as large masses at the time of the diagnosis. So they do not produce symptoms until they grow large enough to compress or invade contagious structures presenting with early satiety. Abdominal discomfort, distension, vomiting and other Gastrointestinal obstructive symptoms [8]. Imaging modalities is of mandatory need for the aid in diagnosing such retroperitoneal sarcomas. It was found that MRI is the preferred mode of use to delineate the extent of the tumor and the relation to adjacent soft tissue and vasculatures. But imaging is not

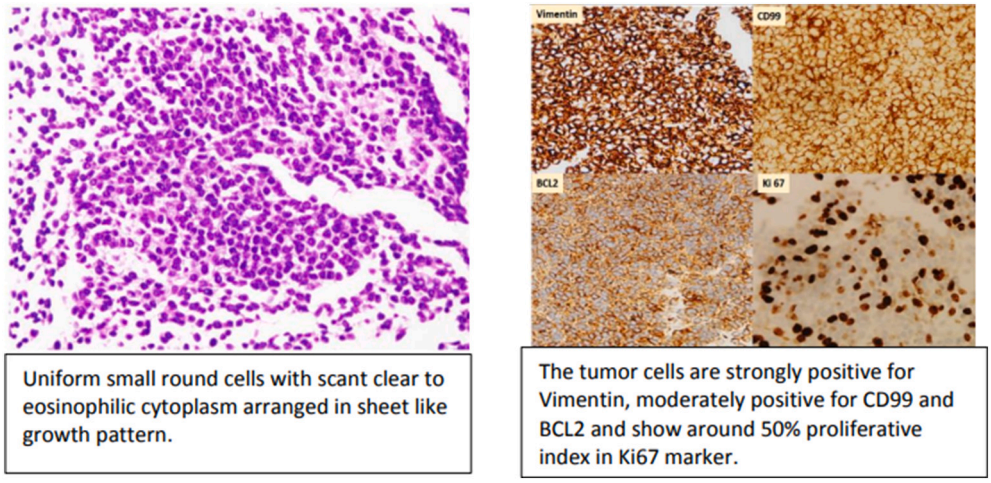

Image 3. Uniform Small round cells with scant clear to eosinophilic cytoplasm arranged in sheet like growth pattern
The tumor cells are strongly positive for Vimentin, moderately positive for CD99 and BCL2 and show around 50% proliferative index in Ki67 marker.

the definitive diagnostic tool due to the expandable features of the retroperitoneal space as it may obscure the details of these masses and add to that there are no specific findings to differentiate abdominal/ retroperitoneal EWS from other equivalent tumors on radiographic images [5]. So it was found that Histomorphology is the definitive diagnostic tool in such tumors [9,10]. In FNAC the tumor cells tend to be uniform and are usually arranged in small tight clusters. The nuclei are usually round and irregular and most of the time it lack nucleoli. It is usually characterized with high nucleocytoplasmic ratio. The cytoplasm is usually pale blue with punched out valocules corresponding to glycogen deposits which will be demonstrated by the acid Schiff stainig – but yet the large amount of intracellular glycogen is not specific for the ES EWS [11]. Homer -Wright rosettes may or may not be present depending on the degree of differentiation [12]. As the histopathological appearance of small round blue cells has an ever expanding list of differentials in case of retroperitoneal EWS depending on the cell lineage such as: 1/Epithelial tumors including small cell Carcinomas, 2/ Mesenchymal tumors as Desmoplastic Small Round Cell Tumors (DSRCTs) and Rhabdomyosarcomas, 3/ Malignant melanoma, and Lymphoma, immunohistochemistry is often mandatory to reach the final diagnosis. Many studies have confirmed that CD99 or MIC which shows membrane positivity was helpful in distinguishing the EWS/PNET group from other MSRCTs [11,13]. Vimentin, NSE and S-100 are also frequently expressed [4], and negative for Desmin and CK [7].

4. Management

ES-EWS is an aggressive tumor with high incidence of local recurrence and distant metastasis that's why is was given its poor prognosis characteristic. Prognostic factors are similar to primary osseous Ewing's Sarcoma, as presence or absence of metastasis, tumor size, extent of necrosis, initial response to chemotherapy and presence of EWS/FL 11 fusin transcripts [14]. Multimodality treatment consisting of adequate surgical resection, aggressive chemotherapy (Vincristine, Doxorubicin, Cyclophosphamide and Actinomycin - D Alternating with Ifosfamide and Etoposide) and high dose-radiotherapy if indicated is recommended [15]. and it has been proven a better survival rate after using Combined [16] and Multidrug [17] Chemotherapy. The strength of this such case is presenting it so more awareness of the condition to be established on the other hand the weakness lies behind its rare incidence and nonspecific signs and symptoms will make the diagnosis to be at a late stage to be intervened properly.

5. Conclusion

Blue Round cell sarcoma is an aggressive type of malignant tumors. Ewing sarcoma is being one of this family group of cancers. It usually presents as Osseous type with rare occasions of extra-skeletal of Ewing's sarcoma. The later with an aggressive course. Yet its pattern is not well studied due to its low incidence and written literature. But it was noticed that it has a good response to combined and multimodal treatment in term of chemotherapy, radiotherapy and aggressive surgical resection.

6. Learning points (what will it add to the literature)

- Early diagnosis of such rare tumor will help targeting it at early stage
- Early diagnosis and management will aid in case being amenable to surgical Curative treatment
- Rare type of tumor that has to have further updates in management

- Patient perspective: patient was aware of his condition and was updated of the full course of management and he was accepting it and appreciating our care to him
- Methodology in writing the Case report: This work has been reported in line with the SCARE 2020 Criteria [18].

Patient consent

Written informed consent was obtained from the patient's father (as the patient passed away at time of taking the consent) for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in Chief of this journal on request.

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Taken from research and research ethics Committee in Bahrain defense force Hospital and form of the Ethical approval is available for review on request.

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CRediT authorship contribution statement

- Fatema Nedham: First author and who Collected data, has written the case report, did the literature review and the discussion
- Veena Nagaraj: Second author, chief resident pathologist who contributed by supplying the Pathology reports and images of the pathology slides
- Abdulla Darwish: Thirds Author, Consultant pathologist, who contributed by supplying the Pathology reports and images of the pathology slides
- Thamer Al-Abbasi: consultant surgery who supervised the whole case and the patient was admitted under his care

Declaration of competing interest

Nil.

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