Case report

Esophageal extraskeletal neoplasm Ewing's sarcoma: Case report

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1. Discussion

Ewing's sarcoma of the paraesophagus is a very unusual condition. Only three cases of esophageal Ewing's sarcoma have been published in the literature to yet, to our knowledge. The majority of occurrences (10–11%) occurred in adults over the age of 20. Monomorphic round cells with small hyperchromatic nuclei, inconspicuous nucleoli, sparse cytoplasm, and large necrotic regions define both skeletal and extraskeletal Ewing’s sarcomas. It's difficult to make a diagnosis for such a rare occurrence. Ewing's sarcoma shares histological and immunophenotypic characteristics with other juvenile small round cell cancers. As a result, an enlarged panel of immunohistochemical tests, fluorescence in situ hybridization, and reverse transcription polymerase chain reaction (RT-PCR) are required to rule out additional diseases such as neuroblastoma, lymphoblastic lymphoma, poorly differentiated synovial sarcoma, and so on [12].

The EWSR1 gene is one of the genes most sensitive to translocation in soft tissue tumors and encodes the EWS protein, which is a member of a growing family of highly conserved RNA-binding proteins mediating interaction with RNA or single-stranded DNA. The codified protein takes part in transcriptional regulation for specific genes and in mRNA splicing. Specifically, EWSR1 is involved in transcription initiation. Concerning EWSR1 breakpoints, the main areas susceptible to breakage are EWSR1 exons 7, 8, 9, or 10 [15].

Fig. 1. Linear cutter stapler gun is used to cut the lesser curvature of esophagus.
Surgery, wherever possible, remains to be the mainstay of treatment [13]. Esophagectomy/esophagogastrectomy is the surgery of choice. Even if metastases are present, a palliative resection can still be performed [13]. Endoscopic resection is another surgical option available [14]. The role of adjuvant radiotherapy and chemotherapy is controversial [13]. Palliative procedures like stenting to relieve dysphagia improve quality of life [16, 17].

1.1. The present case in the context of the literature

Sarcoma is a rare entity among all esophageal malignancies. It presents as an exophytic mass, and in this case, it has presented as a stricture esophagus. Most of these tumors present in locally advanced and disseminated condition, one of the reasons being difficulty and hence delay in diagnosis. In spite of best efforts, a group among them remains to be histologically uncharacterized. Here, we report a case of malignant spindle cell tumor of esophagus, a cause for a stricture esophagus. A definitive histopathological diagnosis could not be achieved.

Even in the case of inoperable disease, palliative resection has a role to play in terms of treatment. The importance of strong local treatment should be highlighted in light of locoregional failure. Endoscopically, polyoid and exophytic masses [18], as well as ulcerating tumors [19], are present. Large intramural masses with ulceration/tracking, expansile intraluminal masses, or areas of luminal constriction may be seen on barium scans [20]. Stricture esophageal stricture is a rare complication. CT/MRI imaging may reveal an intramural mass that is not enhancing uniformly [23]. Submucosal esophageal tumors, which would ordinarily require open biopsy for diagnosis, are one of the criteria for endoscopic ultrasound and its guided biopsy or fine needle aspiration cytology. As a result, the period between diagnosis and treatment may be reduced [21].

2. Case presentation

A 26 years old Asian female referred primarily for surgical treatment due to esophageal cancer detected on her diagnostic investigations and revealed a primary tumor located near the gastroesophageal junction. She was on clinical examination was anemic and her abdominal
examination was unremarkable so she was investigated for her abdominal pain and her CT scan abdomen was done that revealed a heterogeneously enhancing mass lesion seen arising from the lower end of esophagus projecting into the lumen. It measures about 6.3 * 8 cm in AP and transverse dimension, lymph node also noted in lesser sac, appearance are suggestive of lesion involving the GE junction and proximal stomach no pulmonary and hepatic metastases noted.

The PET-CT was performed revealing no bony metastases at the time of scan. She underwent for upper GI endoscopy that showed mass partially obstructing the lumen of esophagus biopsy were taken. Biopsy report came out to be positive for immunohistochemical stains CD99, Cyclin D-1 and NKX 2.2, Ewing sarcoma is a possibility.

Other immunohistochemical stains were performed which showed synaptophysin positive, CD99 positive, NKX2 positive, Cyclin D-1 positive, and cytokeratin CAM 5.2 positive, and interpretation translocation of 23Q-12 is not detected. Based on the results of diagnostic investigations which confirmed the possibility of the tumor Ewing sarcoma of esophagus, she was first followed with neoadjuvant intravenous chemotherapy, after taking three cycles of neoadjuvant chemo showed good response in CT scan and endoscopy the patient underwent McKe-won esophagectomy.

3. Surgical technique

Operation was started with right posterior lateral thoracotomy through the 5th or 6th intercostal space with division of lat dorsi and serratus anterior muscle. The deflated lung is retracted anteriorly for exposure of the posterior mediastinum. The pleura incised and azygous vein divided the esophagus is dissected circumferentially from the level of hiatus into the thoracic inlet. Paraesophageal and subcranial lymph node are incorporated with the specimen to release carefully the gastric tube from adhesions with the right lung and thoracic wall, the esophagus is further dissected bluntly, a chest tube is placed and thoracotomy is closed in layers.

For abdominal phase a supraumbilical incision is made division of falciform and left triangular ligament is made to retract the left lobe of
liver, the right gastric artery is preserved. The abdominal esophagus and nodes dissected, the hiatus opened dissection performed the course of right gastroepiploic artery is determined, the greater curvature of stomach is then mobilized towards the pylorus the left gastric vascular pedicle divided adequate mobilization of stomach done, cervical phase a 6 cm incision was made along the anterior border of left sternocleidomastoid muscle starting at the sternal notch and extending to the cricoid cartilage the platysma and omohyoid muscle divided middle thyroid vein and inferior thyroid vein divide esophagus is further dissected into the superior mediastinum with gentle dissection the esophagus is divide with linear stapler in the neck incision preserving the cervical esophagus.

3.1. Conduit preparation

The stomach and thoracic esophagus delivered out of abdominal incision, lymphatic tissue and right gastric vessel are preserved; starting from the lesser curvature of the stomach the linear cutter stapler is fired towards the fundus of the stomach thus creating a 4–5 cm wide gastric conduit ensuring 5 cm distal to the tumor, the gastric conduit stapler line is then oversewn with a running 3-0 PDS, and the gastric conduit is gently delivered through the mediastinum into the neck.

3.2. Cervical anastomosis

A 45 mm long linear cutting stapler is placed into cervical esophagus and gastric conduit to create posterior wall of anastomosis, an NG tube is placed through the anastomosis under direct visualization. The anterior aspect of anastomosis is completed with 4-0 PDS suture, a penrose drain is placed, the platysma is loosely approximated to sternocleidomastoid muscle with interrupted vicryl suture, the skin is closed. Feeding jejunostomy is located 20 cm distal to duodeno jejunal junction (Figs. 1–5).

Gross pathology showed the tumor to be friable and having multiple ulcerations on the surface. H&E sections revealed a small, blue, round tumor. Histopathological examination showed positive CD99, CK (pan), Ki67 (70 %+), Fil-1, and CD34 levels.

Fig. 4. Cervical esophagus anastomosis.
4. Follow-up and outcome

The patient has been followed up regularly for every 3 month after surgery and for 6 month after adjuvant chemotherapy including clinical examination, ECT, chest CT, and gastroscopy. No obvious signs of recurrence or metastasis were found, and the patient's general condition was satisfactory.

5. Conclusion

In conclusion, we present an uncommon case of extraskeletal Ewing's sarcoma, and discuss its rare presentation and evolution. To our knowledge, this is the first reported case of paraesophageal primary Ewing's sarcoma.

Consent

Written informed consent was obtained from the patient for publication of this case report.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Ethical approval

It's a case report, no ethical approval required for this publication.

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None.

Fig. 5. Removed gross specimen consists of esophagus and fundus of stomach.
CRediT authorship contribution statement

Hina Khalid: For manuscript writing, literature review, interpretation, data collection involve in surgery
Niaz Hussain: Review and analysis
Rafay Shamshad: Review and analysis.

Declaration of competing interest

There are no conflicts of interest.

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1 The work has been reported in line with the SCARE 2020 criteria: Agha RA, Franchi T, Sohrabi C, Mathew G, for the SCARE Group. The SCARE 2020 Guideline: Updating Consensus Surgical CAse REport (SCARE) Guidelines, International Journal of Surgery 2020;84:226–230.