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CASE REPORT OPEN ACCESS

Extraosseous Ewing's Sarcoma Mimicking a Retropharyngeal Abscess: A Rare Case

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1 | Background

Ewing's sarcoma (ES) is an aggressive, small, round blue cell tumor. It is the second most common bone tumor, typically affecting those aged 10–20, with a male preponderance of 1.5:1 [1–3]. ES typically involves the bones, with a higher involvement rate amongst the lower extremities [4]. However, extraskeletal involvement has also been reported [2, 3]. ES is extremely rare in the head and neck region, making up 1%–4% of cases [5]. Of these cases, primary ES, affecting the nasopharyngeal/retropharyngeal area, is even rarer, mainly affecting children [4].

An extensive literature search demonstrated no reported cases of ES in the retropharyngeal area as primary or initial presentation in an adult. Here, we describe the case of a 59-year-old female who was diagnosed with ES in the retropharyngeal area as the initial presentation.

2 | Case Report

A 59-year-old female presented to the emergency department with acute stridor and tachypnoea, requiring 8 L of oxygen. She had a 3-week history of progressively worsening dysphagia and difficulty breathing. She had no other comorbidities, was a non-smoker, and did not consume alcohol.

Comprehensive Ear, Nose & Throat Care (ENT) examination revealed biphasic stridor associated with tachypnoea (respiratory rate 24 breaths/minute), and her oxygen saturations on room air were 90% and required 8 L oxygen. Flexible nasoendoscopy (FNE) demonstrated noticeable laryngeal secretions, indicating an aspiration risk, and a significant posterior pharyngeal wall bulge obstructing the view of the vocal cords (Figure 1). The neck was normal. Inflammatory markers were raised on presentation (white cell count (WCC): 11.3 and C-reactive protein (CRP): 212).

She underwent computed tomography (CT) followed by emergency theatre. CT showed a large retropharyngeal mass, measuring 64 mm craniocaudal, 84 mm transverse and 15 mm in anteroposterior depth. It displaced the larynx anteriorly and effaced the supraglottic airway. Of note, there were bilaterally enlarged level IV nodes up to 21 mm and right level IV nodes up to 20 mm (Figure 2).

Initial emergency surgery included an awake fiberoptic nasotracheal intubation, followed by pharyngolaryngoscopy biopsy and surgical tracheostomy. Pharyngolaryngoscopy demonstrated intraoral diffuse non-ulcerative swelling in the retropharyngeal wall, mimicking a retropharyngeal abscess, which was biopsied. An incision over the swelling demonstrated no pus, and a swab sent for culture was negative for microbiology.

Speech and language therapy assessment revealed profound oropharyngeal dysphagia secondary to the mass, compounded by recent tracheostomy insertion, necessitating nil by mouth status and alternative feeding methods.

An initial diagnosis of pharyngeal pouch with inflammatory changes was suggested. Magnetic resonance imaging (MRI) was performed to gain a better understanding of the nature of the lesion. MRI showed a heterogeneously enhancing, centrally

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necrotic appearing mass, centered on the contiguous masses involving both supraclavicular fossa/level IV. No other convincing abnormal lymph nodes were seen. The vertebra appeared preserved, with no convincing invasion of the vertebra or neural foramina/epidural space (Figure 3). It demonstrated markedly restricted diffusion; lymphoma was considered. Appearances were more suggestive of a neoplasm, possibly originating from the pharyngeal pouch. Additionally, it revealed level IV masses/lymphadenopathy would be amenable to biopsy.

Initial biopsy results showed a poorly differentiated malignant neoplasm composed of sheets of pleomorphic cells with granular chromatin, demonstrating focal nuclear molding and brisk mitotic activity. There are large areas of necrosis and scattered apoptotic debris. Infiltrative is seen focally infiltrating adipose tissue and shows areas of smearing artefact. Occasional tumor giant cells are noted, some of which are multinucleate. Biopsy revealed a highly aggressive, poorly differentiated small round blue cell tumor in the retropharyngeal space with neuroendocrine features. However, subsequent immunohistochemical staining and fluorescence in situ hybridisation (FISH) analysis confirmed the diagnosis as ES, characterized by CD56 and CD99 positivity and an *EWSR1* gene rearrangement.

PET-CT for staging demonstrated vertebral involvement and cervical lymphadenopathy with mesenteric nodules of uncertain significance.

The Head and Neck multidisciplinary team (MDT) meeting confirmed the presence of extensive disease and concluded that treatment should optimize quality of life rather than pursuing curative intent. The proposed chemotherapy regimen consisted of single-agent doxorubicin, administered intravenously every 3 weeks for a maximum of six cycles. If the patient demonstrated adequate response, treatment could be intensified with the addition of consolidation radiotherapy.

During her admission, the disease progressed, necessitating multiple courses of antibiotics for hospital-acquired infections. Despite treatment, the patient's condition continued to deteriorate. Given the advanced disease stage and declining clinical status, including delirium and respiratory distress, the patient's

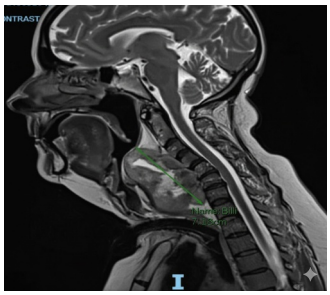

FIGURE 3 | MRI neck showing the large mass in the retropharyngeal area, causing compression.

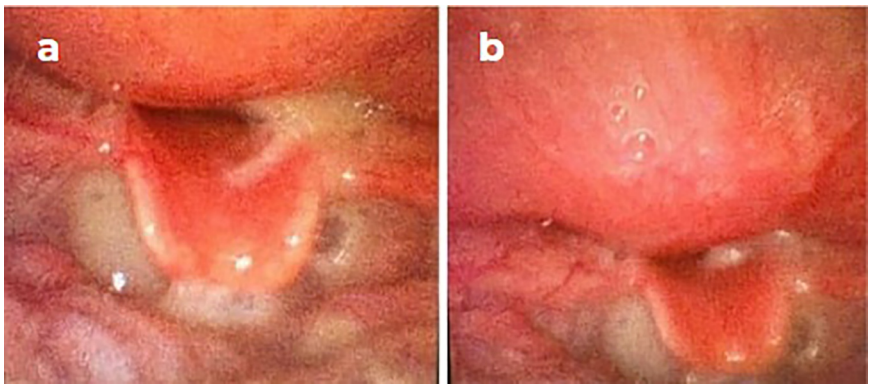

FIGURE 1 | (a and b) Flexible nasoendoscopy showing significant posterior pharyngeal wall bulge obscuring view of vocal cords and noticeable secretions.

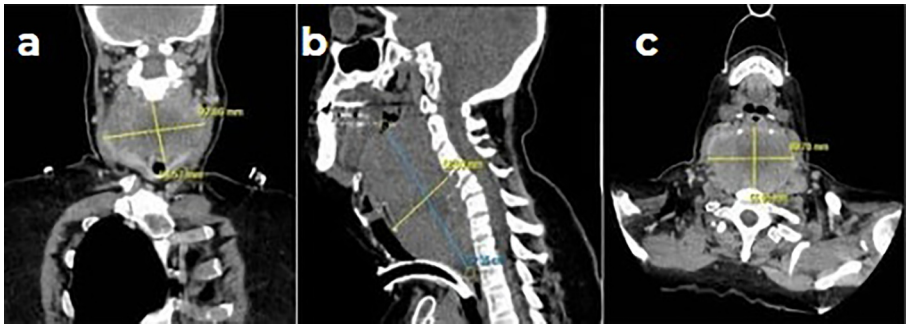

FIGURE 2 | CT neck showing large retropharyngeal mass, displacing the larynx anteriorly and effacing the supraglottic airway. (a) Coronal view. (b) Sagittal view. (c) Axial view.

care transitioned to best supportive care. She subsequently died within 2 months of her initial presentation.

3 | Discussion

The authors describe a unique case presentation of a retropharyngeal ES in an adult which has not been described in the literature. ES comprises 4%–6% of all primary bone tumors. It originates in the marrow cavity and is found in the epiphyses of long and flat bones [5]. The disease most commonly occurs in the second decade with a male preponderance and is rare after the third decade [6].

Common osseous sites of origin include lower extremities (40%–45%), pelvis (20%–25%), chest wall (15%–20%), and upper extremities (10%). Non-osseous tissues of origin include the gastrointestinal tract, kidneys and skeletal muscle. Symptoms vary by site and in 25% of cases, may be attributable to metastases to lungs, bones or bone marrow. Nodal or liver involvement is rare [7].

Involvement of the head and neck in ES is rare, with the skull, vertebrae and mandible being the most commonly affected sites. This could possibly be due to limited haematopoietic marrow in these regions [5, 8]. In the head and neck, swelling and pain are the most common presenting complaints [9]. In this case, the patient presented with symptoms of progressively worsening dysphagia and acute stridor secondary to an enlarging retropharyngeal swelling.

In our case, the extensive, firm mass raised the suspicion of malignancy. Although a retropharyngeal abscess was considered, however the absence of pyrexia, a fluctuant mass and intraoperative pus made this diagnosis less likely. An infected pharyngeal pouch with necrosis was also considered. The diagnosis of ES was challenging, given the rarity of ES in the head and neck region.

Cancer workup should include CT and MRI to characterize the tumor and its relationship with neighboring soft tissue and bony structures, and PET CT considered to assess for distant spread. Biopsy for tissue diagnosis is challenging via fine needle aspirates and usually requires more tissue, making a core or open biopsy preferable.

Typical radiologic features of Ewing's sarcoma include permeative, destructive bone lesions with origin in the metaphysis or diaphysis of long bones [10]. However, the classical signs such as periosteal reaction, cortical thickening, permeative changes, sclerotic change or soft tissue mass are less common in head and neck Ewing's sarcomas [5]. In our case, there were no classical radiological signs of ES, which contributed to the diagnostic challenge.

Treatment is multimodal and should be decided using a multidisciplinary approach. Combination chemotherapy has helped improve survival rates from less than 10% to over 60%. Local treatment may include surgery, radiotherapy, chemotherapy or a combination thereof. For most sites, surgery with adequate margins is the preferred mode of treatment, with radiation given for marginal or intralesional surgery or inoperable tumors [11]. Treatment algorithms for patients with head and neck ES do not significantly differ from those of patients with the more common appendicular ES and include a multimodal approach [12]. However, surgical resection in the case of head and neck ES is not commonly employed due to its risk to surrounding structures and functional outcomes. The size and site of the tumor in the head and neck regions guide the best approach. A negative margin is the gold standard, but it is not possible in case of extensive disease and certain head and neck locations. Similarly, in our case, the extensive mass posed significant challenges to achieving complete resection with negative margins without compromise the surrounding structures or causing significant complications. Given the advanced disease, surgical intervention with curative intent was not feasible [13].

Ultimately, local control is critical to the treatment's success [5, 14]. Therefore, aggressive treatment of the primary tumor is imperative. Chemotherapy is an integral part of therapy since the disease has a propensity for systemic dissemination [15]. Radiation therapy (RT) has been used frequently in these cases since complete ablative surgery is usually not possible due to the difficult location of these tumors [5, 14].

Refined treatment techniques, with the growth of endoscopic transnasal and skull base surgery, may improve oncologic outcomes without risking unacceptable patient morbidity compared to traditional open approaches. Case reports on ES resection with these methods have demonstrated feasibility and success [16, 17]. New RT techniques, such as proton beam RT have been explored for the treatment of tumors in critical locations within the head and neck, such as the skull base [18]. Rombi et al. described the use of proton beam RT in a cohort of 30 patients, four of whom received treatment to the head and neck. The study found similar local control and overall survival to conventional RT, suggesting that with more patients, proton beam RT may reduce local toxicities due to less collateral radiation [19]. Future studies on efficacy and toxicity reduction are needed to confirm the suspected improvements.

ES presenting as a retropharyngeal mass is rare. Definitive diagnosis in these cases can be challenging and should be accompanied by a thorough immunobiological study. MDT approach is required, and treatment options vary. Refined treatment techniques with endoscopic transnasal and skull base surgery may improve outcomes without associated morbidity and new RT techniques such as proton beam RT may reduce local toxicities.

4 | Conclusion

Our case presents a rare instance of ES in the retropharyngeal area. Due to its location and characteristics, diagnosis poses challenges. This report emphasises the importance of considering rare diagnoses in atypical presentations and highlights the necessity of multidisciplinary management in aggressive malignancies. Further research in this area is highly recommended.

Author Contributions

Syed Zohaib Maroof Hussain: conceptualization (equal), data curation (equal), formal analysis (equal), methodology (equal), resources (equal), software (equal), writing – original draft (equal), writing – review and editing (equal). **Priya Dhar:** conceptualization (equal), data curation (equal), formal analysis (equal), software (equal), writing – original draft (equal), writing – review and editing (equal). **Madeleine Grand:** conceptualization (equal), methodology (equal), writing – original draft (equal), writing – review and editing (equal). **Charles Hall:** conceptualization (equal), data curation (equal), methodology (equal), supervision (equal), writing – original draft (equal), writing – review and editing (equal). **Declan Costello:** formal analysis (equal), supervision (equal), writing – original draft (equal), writing – review and editing (equal).

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Ethics Statement

The authors have nothing to report.

Conflicts of Interest

The authors declare no conflicts of interest.

Data Availability Statement

All relevant data is viable upon request.

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