CASE REPORT

A Rare Presentation of Synovial Sarcoma as Cervical Lymphadenopathy in a Pediatric Patient—a Case Report

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Abstract Synovial sarcoma is the uncommon malignant tumor of children and adolescents. It usually involves the soft tissues of the upper and lower joints and most commonly affects adults in their fourth decade of life. We report a rare case of synovial sarcoma of the head and neck region in a five-year-old child who has presented with right side cervical lymphadenopathy associated with dysphagia and hoarseness of voice. Patient was managed by modified radical neck dissection followed by chemotherapy.

Keywords Synovial sarcoma · Pediatric tumor · Neck dissection · Cervical lymphadenopathy

Introduction

Sarcomas are malignant mesenchymal soft tissue tumors representing only 1% of all malignant tumors, about 8% of

all pediatric cancers and 1% of all head and neck malignancies [1]. Synovial cell sarcoma (SS) is a high-grade histological variety and it is the fourth most common entity. It mainly occurs in young adults with a male/female ratio of 2:1. It is mainly located near large joints and bursae of the lower extremities such as the knee, tendon sheaths, and bursal structures. Its occurrence is rare in the head and neck region. Within the head and neck region, common sites are the hypopharynx and parapharyngeal spaces. It is reported that synovial sarcoma bears better prognosis in childhood as compared to adulthood [2]. Due to the rarity of tumor, very few cases have been reported and treatment is still not standardized.

Case Report

A five-year-old male child reported to our hospital with complaints of right-sided neck swelling associated with cough, dysphagia, and hoarseness of voice since the last 3 months. Physical examination revealed enlarged cervical lymph nodes at Level II of size 5 × 3 cm (Fig. 1) which were firm in consistency, non-tender, and fixed to the underlying tissue. There was no local rise in temperature and the overlying skin was normal. Sonography of neck revealed a large heterogeneous mass of size 4.2 cm on the right side with multiple cervical lymphadenopathies. Trucut biopsy reported malignant round cell tumor. Immunohistochemical analysis further confirmed the diagnosis of poorly differentiated malignant round cell tumor possibly synovial sarcoma. Positron emission tomography (PET) scan was done to exclude metastasis to distant organs which (Fig. 2) revealed two metabolically active, slightly enhancing soft-tissue density lesions in the right upper jugular, mid-jugular, and posterior cervical space extending to the carotid space suggesting primary neoplastic/metastatic lymph node lesion (SUV max 3.6). Modified

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Fig. 1 Intraoperative view

radical neck dissection was done and we found that the tumor was arising near the joint space of the mandible on its medial surface. Tumor was excised without mandibular split technique as it was delivered with retraction only. On histopathological examination of neck dissection, 8 out of 22 lymph nodes were positive for metastasis. The child was subjected to six cycles of chemotherapy (ifosfamide + etoposide

alternately with vincristin + doxorubicin + endoxan) and the disease responded well. Patient was on follow-up and is doing well till six monthly follow-up.

Discussion

Synovial sarcoma is a malignant high-grade soft tissue tumor having high risk of local invasion and metastasis. It can occur at all ages but most frequently affects young adults and adolescents. It is rarely reported in children of age 4–8 years. Synovial sarcomas do not originate from synovial tissues, but from pluripotent mesenchymal cells which may be close or remote from the articular surfaces. In the head and neck, the hypopharynx is the commonly involved site. The hypopharynx may also be the most frequent site of head and neck synovial sarcoma because it is the seat of numerous synovial formations. Other locations in the head and neck reported in the literature include the masticator space, parapharyngeal space, sinonasal region, and pharynx [2, 3]. Synovial sarcomas have also been found in unusual sites in the oral and maxillofacial region such as the cheek mucosa [4].

The prognosis depends mainly on the tumor size, proximity to adjacent vital structures, and presence of any metastasis.

These tumors are chemo sensitive and response rates are high in pediatric tumors. [5–7] Chemotherapy with ifosfamide is successful mainly for synovial sarcoma of the hypopharynx and larynx where complete excision is sometimes not possible. The multimodal treatment of surgical resection with adjuvant radiotherapy or/and chemotherapy has improved the prognosis of synovial sarcoma in children. The ideal surgical approach is wide-local excision to achieve negative surgical margins for improved local control as well as longer overall survival. However, negative margins can be difficult to achieve mainly in the head and neck region due to limited space and proximity to vital structures. There is no indication for prophylactic neck dissection since there is no lymphatic spread within the natural course of synovial sarcoma. But in case of palpable neck nodes, neck dissection is indicated. Foreman et al. reported local recurrence rates of up to 60–90% following inadequate surgical resection without the use of adjuvant radiotherapy. [8] Though no standard radiation treatment regimen exists, radiation doses of 60–70 Gy are commonly used. Harb et al. reported that patients treated with surgery and adjuvant radiotherapy had lower recurrence rates and higher disease-specific and overall survival rates as compared to those treated with surgery alone, regardless of margin status. [9]

In the literature review of past 10 years, studies have found recurrence rates ranging from about 20 to 60%. As distant metastasis is common with synovial sarcoma of the head and neck, effective systemic therapies should be evaluated in order to improve survival and quality of life.

Compliance with Ethical Standards

Conflict of Interest All the authors declare that they have no conflict of interest.

Ethical Approval This article does not contain any studies with human participants or animals performed by any of the authors.

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