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ORIGINAL ARTICLE

A Case Series on Unusual Neck Masses

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Abstract Neck masses can be defined as any abnormal swelling or growth from the level of base of skull to clavicle. They can be benign or malignant so a thorough investigation is necessary to reach to a final diagnosis. Here we report a case series of three unusual neck masses presenting to the Out patient Department of Otorhinolaryngology and Head and Neck Surgery in R. G. Kar Medical College, a tertiary care hospital of Kolkata in a span of 1.5 years. The rarity of the etiology behind the neck masses makes this case series unique.

Keywords Plasmacytoma · Sarcoma · Lymphangioma · Excision

Introduction

Neck masses are often seen in clinical practice, and it is very vital to determine the etiology of the mass using efficient diagnostic methods. The primary goal is to determine if the mass is malignant or benign. If the history and physical examination donot bring us to a conclusive diagnosis, imaging and surgical tools are used. Contrast-enhanced computed tomography is the initial diagnostic test of choice in adults. Computed tomography or a Magnetic Resonance imaging is usually done. A fine-needle aspiration biopsy provides diagnostic information via cytology, Gram stain, and bacterial and acid-fast bacilli cultures.

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Primary plasmacytoma of lymph nodes is very rare. It represents 2% of all extramedullary plasmacytomas, and only 0.08% of all plasma cell malignant neoplasms [1] Primary plasmacytomas usually present as solitary lesions of the bone [2] An extramedullary plasmacytoma is a plasma cell neoplasm of the soft tissue without involvement of the bone marrow; most common being of the upper aerodigestive tract [3]. In case of extramedullary primary plasmacytoma of lymph nodes, there is abnormal monoclonal proliferation of plasma cell in the tumour of lymph nodes in absence of any systemic involvement. Most primary plasmacytomas of the lymph nodes are seen in the cervical lymph nodes, usually presenting as neck masses. These tumours are extremely rare with less than 40 cases described in the literature till date [4].

Ewing sarcoma is a poorly differentiated round cell tumour with high malignant potential. They are usually osseus in origin however, rare but may have an extraosseus presentation as well. These are very aggressive tumours with high rate of local recurrence and distant metastasis. Commonly affected extraskeletal sites include the paravertebral spaces, lower extremities, head and neck and pelvis [5].

Cervical lymphangiomas are uncommon benign congenital lymphatic malformation usually present in children and rare in adults. Lymphangiomas are malformations of the lymphatic system characterized by lesions that are thin-walled cysts; these cysts can be macroscopic or microscopic [6] Acquired lymphangiomas may result from trauma, inflammation, or lymphatic obstruction. Most lymphangiomas are benign lesions that result only in a soft, slow-growing, "doughy" mass. No malignant potential present.

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Case 1

A 62 year old male, presented to the ENT OPD with a history of gradually progressive, painless swelling on the right side of neck for 3 years. Clinical examination revealed multiple swellings involving level II, III, IV, V on the right side of neck, approximately 6 × 5 cm which were firm, non-tender, mobile, without any erythema or ulceration of the overlying skin. Search for similar nodes elsewhere in the body was all for naught (Fig. 1).

Fibre Optic Laryngoscopy and Diagnostic Nasal Endoscopy were non contributory. FNAC performed taking aspirate from 2 sites, viz. anterior and posterior triangle nodes and was suggestive of Plasmacytoma. HRSG neck and CECT neck were suggestive of right sided cervical lymphadenopathy largest one being 48 × 40 mm (Fig. 2). Incisional Biopsy was performed from the right cervical mass under local anaesthesia and histopathological report clinched the diagnosis of Plasmacytoma. A full work-up was conducted to search for any other evidence of plasma cell dyscrasia. Initial laboratory investigations revealed Hb-13.4 g/dl, Sodium-137 mg/dl, Urea-17 mg/dl, Potassium-4.6 mg/dl, Creatinine-0.89 mg/dl, Calcium-9.4 mg/dl. β2-microglobulin- 1600mcg/l. Bone Marrow Biopsy showed uninvolved marrow. Whole Body PET CT Scan showed FDG avidity in only right cervical nodes II-V, largest being in the level II (4.7 × 3.9 cm). Serum protein electrophoresis (SEP) didnot yield a monoclonal spike (M spike) in the gamma globulin region. Serum immunofixation electrophoresis didnot reveal any monoclonal gammopathy. Bence Jones protein was absent in urine protein electrophoresis. A skeletal survey was conducted which included a X ray of the skull, long bones,and axial skeleton. The skeletal evaluation revealed no osteolytic lesions.

So, from the above findings we can easily exclude mutilple myeloma as we see an uninvolved bone marrow, no M spike on SEP and no evidence of end organ damage i.e. CRAB criteria.

Hence, a final diagnosis *of primary extramedullary plasmacytoma* of cervical lymph node was made.

The patient was treated via radiotherapy,a total dose of 50 Gy in 25 fractions. On treatment, there was complete resolution of the mass and the patient was followed up at 3 month interval for a period of 1 year and no recurrence was noted.

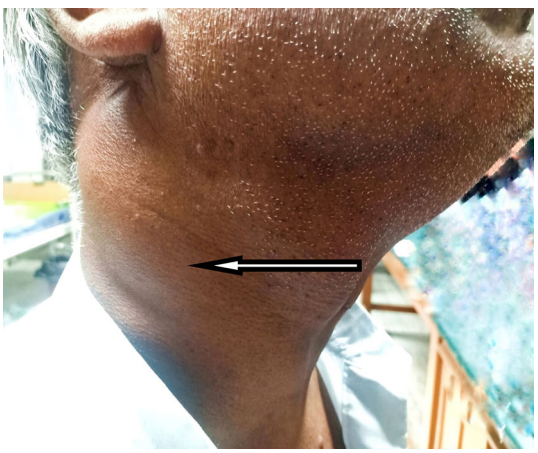

Fig. 1 Right sided neck swelling involving level II, III, IV, V

Case 2

A 11 year old female presented to the Outpatient Department with swelling on the left side of the neck for 8 months (Fig. 3). It was associated with intermittent episodes of pain over the swelling and dysphagia with a history of weight loss. No history of fever, trauma, night sweat, dyspneoa and cough. No contact history was present.

On Examination,a swelling with approximate size 5 × 4 cm was palpable involving level Ib, II, III and V group of cervical lymph nodes. It was firm in consistency,smooth surface,regular margin, mobile,transilluminant, fluctuant, non tender without ulceration of the overlying skin. On Fibreoptic Laryngoscopy, a smooth mass was seen protruding through the left tonsillar fossa pushing the left anterior tonsillar pillar medially. Larynx and base of tongue was normal with no compromisation of airway (Fig. 4). MRI Neck showed a large lobulated altered signal intensity is seen at the left parapharyngeal space extending to retropharyngeal space without obliteration of

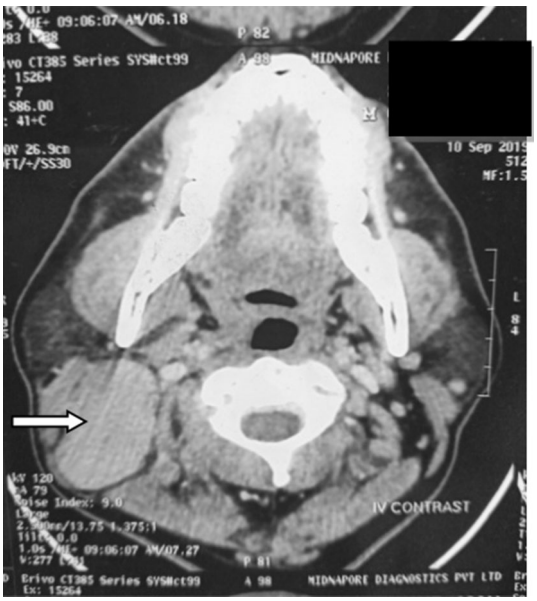

Fig. 2 CECT neck showing right sided cervical lymphadenopathy, largest being 48 × 40 mm

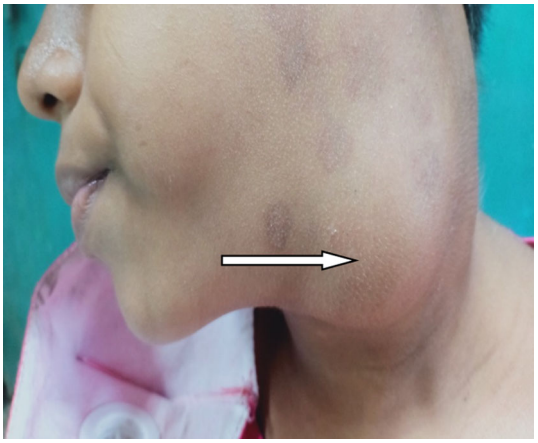

Fig. 3 Left sided neck swelling involving level Ib, II, III, V

fat plane between deep lobe of parotid gland and the mass. FNAC was suggestive of features of round cell neoplasm.

Surgical excision via transcervical approach was planned under general anaesthesia and the specimen was sent for histopathological examination and immunohistochemistry. The postoperative period was uneventful.The result revealed it to be an ***Extraosseus Ewing Sarcoma***. The patient is lost to follow up because of the ongoing COVID 19 situation.

Case 3

A 24 year old female presented to the Outpatient Department with a swelling on the right side of neck for a duration of 1.5 years. It was associated with no other symptoms

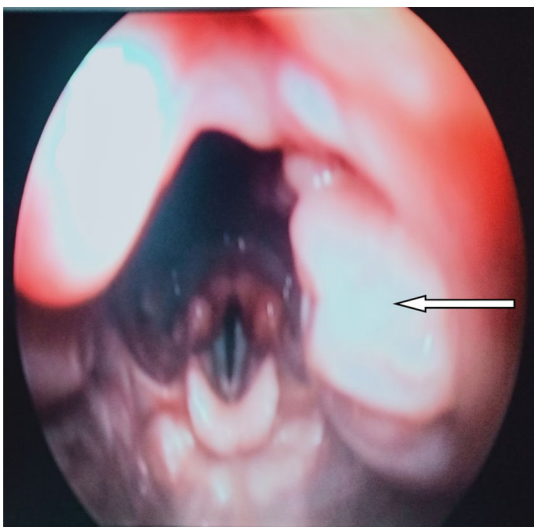

Fig. 4 Mass seen coming through the left tonsillar fossa on fibreoptic laryngoscopy with larynx being normal

(Fig. 5). There was no history of trauma. No history of similar swelling in any other part of the body. On examination,approximately 4 × 3 cm globular swelling was seen on the right submandibular region which was gradually increasing in size, non tender, cystic, smooth margin, non fluctuant, non transilluminant, and not fixed to surrounding structure.There was no erythema or ulceration or venous prominence overlying the cystic swelling.

USG neck showed a 3.9 × 2.3 × 4 cm multiseptated cystic lesion on the right side of neck abutting the deep lobe of parotid and submandibular gland. Right sided neck vessels were normal. Features were suggestive of lymphangioma. CECT neck was done which showed a cystic (H.U. + 3 to + 7) space occupying lesion of 21*28 mm in size with lobulated margin at posterior aspect of right parotid gland extending downwards upto right submandibular region. The tissue plane between parotid gland and the lesion was well maintained. Carotid sheath and its contents were normal. On FNAC, smears showed plenty of small lymphocytes, cyst, macrophages on a thin proteinaceous background. Features were suggestive of lymphangioma.

Patient was planned for surgery (*Transcervical excision of the neck mass under general anaesthesia)* (Fig. 6). Under general anaesthesia, after proper positiong and antiseptic dressing and draping, swelling was marked, 1:100,000 adrenaline was infiltrated along the incision line and incision was given 2 fingerbreadth below the body of mandible along the Langer's line with 15 no. blade. Subplatysmal flap was raised, sternocleiodmastoid muscle was retracted, posterior belly of digastric muscle was delineated,cystic swelling in the right submandibular region was delineated and excised in toto and was sent for histopathological examination. A corrugated rubber drain was placed and wound was closed in layers. The postoperative period was uneventful. Drain was removed after 48 h and stitches were removed after 7 days. Patient was

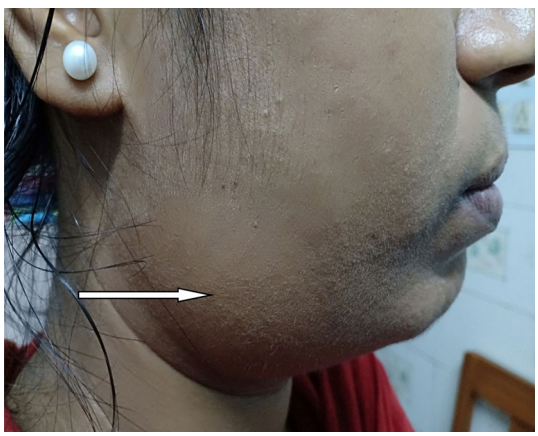

Fig. 5 Right sided neck swelling involving submandibular region

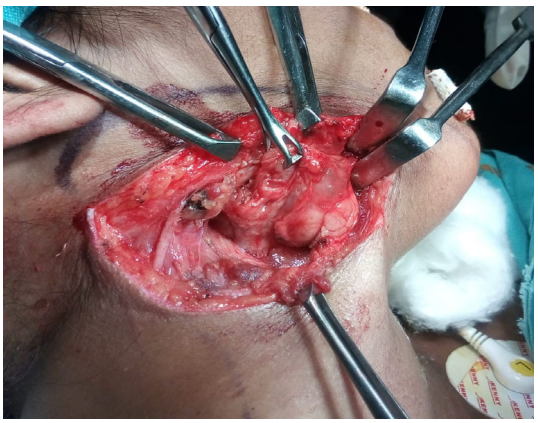

Fig. 6 Transcervical excision of left sided lymphangioma under general anaesthesia

doing well in subsequent follow ups. Histopathology revealed small and large communicating cysts with cyst wall containing lymphoid aggregates with a diagnosis of ***cystic lymphangioma.***

Discussion

Solitary plasmacytomas are uncommon neoplasms, which may manifest as solitary plasmacytoma of the bone, solitary extramedullary (extra-osseous) plasmacytoma or multiple solitary plasmacytomas. Primary extramedullary plasmacytomas are uncommon and rarely progresses to multiple myeloma or plasma cell leukaemia as compared to osseous plasmacytomas [7, 8]. In order to diagnose primary plasmacytoma of the lymph nodes, there must not only be no evidence of plasma cell proliferation elsewhere, but also no associated malignant lymphoma components. As these tumours are extremely radiosensitive so radiotherapy is the treatment of choice and not surgical excision.

Round cell neoplasms are described as round cells with undifferentiated tumour cells. They include Ewing sarcoma, Neuroblastoma, Rhabdomyosarcoma, Peripheral Neuroectodermal tumour, Non Hodgkin's lymphoma etc. Extraosseous Ewing Sarcoma has no specific clinical picture. The most frequently presenting symptom is a rapidly growing mass with local pain [9]. However, the presentation of tumours depends on the sarcoma's site of origin. Imaging also doesn't guide to a definite diagnosis thus the imaging findings being non specific in nature. However, fine needle aspiration cytology can throw some light and histopathological examination, being, confirmatory for the diagnosis. These are quite radiosensitive, but improvements in surgical technique and the risks associated with radiation (secondary malignancies) have reduced the reliance upon radiation [10].

Lymphangiomas are classified as lymphangioma simplex, cavernous lymphangioma and cystic hygroma [11] It is well recognised in paediatric practice but seldom presents de novo in adulthood. [12] The most common sites are in the posterior triangle of the neck (75%), axilla (20%), mediastinum (5%), groin, retroperitoneal space and pelvis. In adults, lymphangiomas may occur either spontaneously or in response to infection or trauma. [13] Majority of cervical lymphangiomas in adulthood are asymptomatic, have no gender predilection and present as a painless mass that enlarges progressively. The lesion is soft, nontender, transilluminant, fluctuant and fixed to deep tissues, similar to pediatric presentation [14] Various differential diagnoses like thymic cyst, pericardial cyst, bronchogenic cyst, cystic teratoma and cystic thymoma, should be kept in mind. The treatment of choice is surgical excision and there is a 15% recurrence rate if the lesion is not fully excised [15].

Conclusion

Neck mass can be the presentation of various diseases, benign or malignant or metastatic. Hence, a thorough detailed physical examination and diagnostic methods is of utmost importance to reach to a final definite diagnosis without delay so that a prompt management of the disease can be started at the earliest. In this case series we came across 3 such unusual causes of neck mass which are very rare and often misdiagnosed. Depending on the nature of the disease the best treatment option needs to be executed to have a long term disease free survival without recurrence.

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