CASE REPORT

SYNOVIAL SARCOMA OF THE LARYNX

Nabila Javed, Javed Iqbal
Institute of Radiotherapy and Nuclear Medicine (IRNUM), Peshawar-Pakistan

Synovial sarcoma is a mesenchymal spindle cell tumour that displays variable epithelial differentiation. It most commonly occurs in lower extremities. Head and neck is a rare site for synovial sarcoma accounting for less than 10%. Larynx is an extremely rare site and only 16 cases with laryngeal location have been reported. Immunohistochemistry is important for correct diagnosis. Surgical excision of the tumour with clear margins and local radiotherapy is effective in local control. Chemotherapy is indicated in the presence of distant metastasis. Case of a 16 years old female is presented with hoarseness of voice and mass in supraglottic region. Lateral pharangotomy and excision of mass revealed synovial sarcoma. She had been treated with adjuvant radiotherapy in September 2012. She was fine and coming for regular follow up.

Keywords: Synovial sarcoma, Larynx

J Ayub Med Coll Abbottabad 2015;27(3):729–30

INTRODUCTION

Synovial sarcoma is a mesenchymal malignancy of unknown histogenesis. Over 80% of cases arise in deep tissues of extremities with 50% in lower extremities. It may be encountered in regions without apparent relationship to synovial structures including the head and neck in less than 10% of cases[1], thoracic and abdominal wall (fewer than 10%). Of the large and heterogeneous group of soft tissue tumours angiosarcoma, epitheloid sarcomas, fibrosarcomas, leiomyosarcomas, malignant fibrous histocytomas, liposarcomas, and rhabdomyosarcomas have been reported in the head and neck. Laryngeal location is extremely rare for synovial sarcoma. Only 16 cases have been reported up till now in literature. Most of the cases were treated either with surgery alone, or combined with radiotherapy. Only two cases were reported in whom chemotherapy was administered.

CASE REPORT

A 16 years old female patient presented with pain in throat of three months duration and hoarseness of voice for two months to ENT department of Khyber Teaching Hospital Peshawar. Indirect laryngoscopy showed mass in the supraglottic region. Lateral pharangotomy and excision of mass was done. Histopathological examination showed spindle cell neoplasm. Size of the mass was 5×4×3 cm with irregular external surface. Immunohistochemical stains were applied that confirmed the nature of malignancy that was synovial sarcoma. Her postoperative contrast enhanced computerised tomography showed focal asymmetrical thickening of mucosa along the left anterolateral wall of supraglottic region in close relationship of tip of epiglottis. No cervical lymphadenopathy was seen. Metastatic work up for distant metastasis was negative. She was started on radiation therapy. She was planned for 60 Gy external radiation on 6 MV photon by two parallel opposed lateral fields to neck. For the first phase 40 Gy was delivered and after 40 Gy, field was reduced to exclude the spinal cord from the field in the second phase. Patient was examined weekly for any side effects of radiation and was managed accordingly.

DISCUSSION

Synovial sarcoma accounts for 5–10% of all soft tissue sarcomas.[1–3] It may be diagnosed at any age but the majority of cases occur in young adults between 15–35 years of age and more commonly in males. Over 80% arise in deep soft tissues of extremities with 50% cases in lower limb and remainder in upper limb. Head and neck is a rare (3–9%) but well known location of synovial sarcoma. Only sixteen cases of laryngeal sarcomas have been reported up till now.[2,4,8,16] It may also be encountered in region without apparent relationship to synovial structures including thoracic and abdominal wall or intrathorasicsites. It generally does not originate from synovial tissue. Case of a 16 years old female patient was presented with supraglottic tumour measuring about 5×3×4 cm as in most of the reported cases. The patient age is typical for synovial sarcoma which is the tumour of younger age group.

Primary laryngeal tumours are predominantly Squamous cell carcinoma.[4] Other unusual tumours in this location are fibrosarcoma, chondrosarcoma, osteosarcoma and rhabdomyosarcoma.[2,5] Histologically it may be monophasic or biphasic (composed of morphologically two distinct types of cells), with a characteristic pattern of epithelial cells surrounded by spindle or fibrous component. Synovial sarcoma in our case was monophasic. Calcification with or without ossification is seen in 10% of tumours, and synovial sarcoma may be confused with other calcifying tumours e.g., synovial sarcoma in lower neck need to be distinguished from thyroid neoplasm,

which may also exhibit calcification. The spindle cells stain positive for keratin and epithelial membrane antigen. Vimentin is demonstrable in spindle cells but absent in epithelial cells. In this case Bcl2, vimentin and CD99 was positive. Nearly all synovial sarcomas contain a characteristic translocation, t(X; 18) (p11.2; q11.2) 144 D fusing the SS18 (SYT) gene with either SSX1, SSX2or SSX4. Hundred percent of biphasic and 96% of monophasic synovial sarcomas possess this translocation. It has become the gold standard in diagnosing synovial sarcoma. Cytogenetic studies were not provided in histopathology report in our case.

Local treatment follows the general principles of soft tissue sarcoma treatment with adequate excision and adjuvant radiotherapy when appropriate, with or without adjuvant chemotherapy.[1,3,4] Local recurrences of up to 80% have been reported after inadequate surgery without radiotherapy.[3] Lymph node dissection is not necessary in most cases[2,4] as there was mass only in suraglottic region so only excision of mass was done. Neck dissection was not done in the presented case. The reported cases in the literature got local radiotherapy in most of the cases, only three cases underwent chemotherapy to whom ifosfamide was given in high doses.[2,6,7] In our case only loco-regional radiotherapy was given after excision of the mass. Negative prognostic factors that imply a high risk of distant metastasis are capsular invasion, age older than 25 years, tumour size more than 5 cm, and poor differentiation of the tumour cells. A proliferation index evaluated by Ki-67 immunostain, higher than 10% is associated with dismal prognosis.[7]

In conclusion synovial sarcoma of head and neck should be considered an aggressive tumour like its counterpart in limbs. Here we are going to add new case of laryngeal sarcoma in the literature. Because of limited number of cases in the literature every new case may bring new information about treatment.

REFERENCES

1. Weiss SW. Goldblum JR. Malignant soft tissue tumours of uncertain type.In: Enzinger and Weiss's Soft Tissue tumors.4thed. St Louis Mo Mosby 2001. Dei Tos AP, Dal Cin P, Sciot R, Furlanetto A, Da Mosto MC, Giannini C, *et al*. Synovial sarcoma of the larynx and hypopharynx. Ann Otol Rhinol Laryngol 1998;107(12):1080–5.
2. Skytting B. Synovial sarcoma. A Scandinavian sarcoma Group Project. Acta Orthop Scand Suppl 2000;291:1–28.
3. Boniver V, Moreao P, Lefebvre P. Synovial Sarcoma of Larynx: Case report and literature review. B–ENT 2005;1(1):47–51.
4. Garensteina, Neel HB 3rd, Weiland LH, Devine KD. Sarcomas of the larynx. Arch Otolaryngol 1980;106(1):8–12.
5. Morland B, Cox G, Randall C, Ramsay A, Radford M. Synovial sarcoma of the larynx in a child;case report and histological appreances. Med Pediatr Oncol 1994;23(1):64–8.
6. Bilgic B, Mete O, Oztürk SA, Demiryont M, Keles N, Basaran M. Synovial sarcoma :a rare tumor of Larynx. Pathol Oncol Res 2003;9(4):242–5.
7. Ferlito A, Caruso G. Endolaryngeal synovial sarcoma. An update on diagnosis and treatment. ORL J Otorhinolaryngol Relat Spec 1991;53(2):116–9.
8. Gatti WM, Strom CG, Orfei E. Synovial sarcoma of laryngopharynx. Arch Otolaryngol 1975;101(10):633–6.
9. Geahchan NE, Lambert J, Micheau C, Richard JM. [Malignant synovioma of the larynx] .Ann Otolaryngol Chir Cervicofac 1983;100(1):61–5.
10. Kitsmaniuk ZD, Volkon IuM, Demochko VB, Ivanov AD. [Synovial sarcoma of the larynx]. Vestn Otorinolaringol 1985;(2):61–2.
11. Capelli M, Bertino G, Morbini P, Proh M, Falco CE, Benazzo M. Co2 Lazer in the treatment of Laryngeal Synovial Sarcoma: A Clinical Case .Tumori 2007;93(3):296–9.
12. Miller LH, Santaella-Latimer L, Miller T. Synovial sarcoma of the larynx. Trans Sect Otolaryngol Am Acad Ophtalmol Otolaryngol 1975;80(5):448–51.
13. Pruszczynski M, Manni JJ, Smedts F. Endolaryngeal synovial sarcoma: Case report with immunohistochemical studies. Head Neck 1994;11(1):76–80.
14. Quinn HJ Jr. Synivial sarcoma of the larynx treated by partial laryngectomy. laryngoscope 1984;94(9):1158–61.
15. Al-Nemer A, EI-Shawarby MA. Laryngeal synovial sarcoma: Case report and Literature review. Gulf J Oncolog 2011;(9):52–6.

Address for Correspondence:

Dr Nabila Javed, Consultant Oncologist, Institute of Radiotherapy and Nuclear Medicine (IRNUM) Peshawar-Pakistan
Ph: +92 91 9216114-7
Email: drnabila67@yahoo.com