A Rare Case of Giant Cell Tumor of Bone with Thoracic Extension with Horner's Syndrome

Saswata Ghosh, Susmita Kundu, Saurav Kar, Palash Nandan Dhara

ABSTRACT

Giant-cell tumor of the bone (GCTOB) generally accounts for 4-5% of primary bone tumors and ~20% of benign bone tumors. It is characterized by the presence of multinucleated giant cells (osteoclast-like cells). Although classified as a benign tumor, GCTOB has been observed to metastasize to the lungs in up to 5% of cases. Patients usually present with pain and limited range of motion caused by tumor's proximity to the joint space. There may be swelling as well, if the tumor has been growing for a long time. Some patients may be asymptomatic until they develop a pathologic fracture at the site of the tumor. The symptoms may include muscular aches and pains in arms, legs and abdominal pain. Patients may also experience nerve pain which feels like an electric shock. In a population based study of Horner’s syndrome in the pediatric age group, the incidence of Horner’s syndrome was estimated to be 1.42 per 100 000 patients younger than 19 years, with a birth prevalence of 1 in 6250 for those with a congenital onset. We are presenting a case of a 27 year old male patient who presents with right sided chest pain and cough for 3 months and absence of perspiration of 2 months which was ultimately proved to be a case of Giant cell tumor of bone with thoracic extension with Horner’s syndrome.

Key words: Giant cell tumor of bone; Horner’s syndrome

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CASE HISTORY

A 27 year old male patient (Body length 1.55 meter, body weight 70 kg) from a suburban area of Kolkata, India came to us (on 23.4.2013) with the chief complaints of right sided chest pain and cough for 3 months and absence of perspiration on the right side of the body for 2 months. Right sided chest pain was both at the front and the back with insidious onset and gradually increasing severity, enough to disturb his sleep and day to day activities.
There was radiation of the pain along inner border of right arm and forearm. Cough was dry in nature with scanty mucoid expectoration and occasional nocturnal exacerbation. There was no episode of haemoptysis as well. Regarding family history, none of his family members have any history of neoplasm and there was no significant history of taking medications of his mother while she was pregnant. Regarding personal history, patient is non-smoker, non-alcoholic, farmer by occupation and not exposed to any chemical substance. On general survey, patient had grade 2 clubbing and no peripheral lymphadenopathy.

On examination of the respiratory system, dull percussion note on along right midclavicular line from 2nd Space downwards and diminished VBS on right infraclavicular, mammary and inframammary area were noted. Evidence of Horner’s syndrome i.e. ptosis, miosis, anhydrosis, enophthalmos and loss of cillio-spinal reflex (Figure 1) was observed along with atrophy of right hypothenar eminence, diminished power of right biceps with exaggerated right elbow and wrist jerk. Blood report shows RBC 4.5 l/cu mm and WBC 8000/cu mm. His chest x-ray postero-anterior view (on 30.4.2013) showed homogenous opacity occupying upper and mid zone of right hemithorax (Figure 2) which was further been corroborated by his CT thorax images (Figure 3 A and B on 03.5.2013). CT guided fine needle aspiration cytology report (on 10.5.2013) showed acute on chronic organizing inflammation with epithelial atypia and giant cell reaction. Subsequently we went for tru-cut biopsy which revealed soft tissue extension of giant cell tumour of bone (on 20.5.2013). Detailed histopathology described that the Tumor was composed of round, oval or spindle shaped cells showing mild to moderate nuclear pleomorphism and diffusely distributed multinucleated osteoclast like giant cells (Figure 4). Immunohistochemistry (on 30.5.2013) shows that the tumour is cytonegative for cytokeratin, epithelial membrane antigen (EMA) and thyroid transcription factor-1 (TTF-1) (Figure 5). Patient was further referred to CTVS department where surgical excision of the tumor was done (on 14.6.2014) and post-operative radiotherapy was given (50 gy in 25 fractions i.e. 5 days a week over 5 weeks). Patient is relatively stable and asymptomatic at present but features of Horner syndrome still persists.
Lateral medullary syndrome.
Cluster headache - combination termed Horton’s headache.
Trauma-base of neck, usually blunt trauma, sometimes surgery.
Middle ear infection.
Tumors-often bronchogenic carcinoma of the superior fissure (Pancoast tumor) on apex of lung.
The diagnosis of giant cell tumors is based on biopsy findings. The key histomorphologic feature is, as the name of the entity suggests, (multinucleated) giant cells with up to a hundred nuclei that have prominent nucleoli. Surrounding mononuclear and small multinucleated cells have nuclei similar to those in the giant cells; this distinguishes the lesion from other osteogenic lesions which commonly have (benign) osteoclast-type giant cells.
But the thoracic extension of the giant-cell tumor of the bone is extremely rare and that presenting with Horner’s syndrome is an extremely unusual condition.

CONFLICT OF INTERESTS
There are no conflicts of interest with regard to the present study.

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Peer reviewers: Xuewu Zhang, Professor, College of Light Industry and Food Science, South China University of Technology, 381 Wushan Road, Guangzhou 510640, China; Ashraf K Khalil, Professor, Institute of Biotechnology & Genetic Engineering, City for Scientific Research & Technology Applications, Alexandria, 21934, Egypt.