Case Report

Aggressive Fibromatosis of Neck and Superior Mediastinum: A Case Report

Cheerasook Chongkolwatana, MD[1], Siriporn Limviriyakul MD[1],
Worawong Slisatkorn MD[2], Warut Pongsapich, MD[1]

[1] Department of Otorhinolaryngology, Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok, Thailand
[2] Division of Cardio-Thoracic Surgery, Department of Surgery, Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok, Thailand

Background: Aggressive fibromatosis is rare in the neck and superior mediastinal area

Case Report: A 76-year-old woman initially presented with a large anterior neck mass with upper airway obstruction due to tracheal compression. Initial biopsy report suggested follicular carcinoma, but final diagnosis was aggressive fibromatosis. Surgical excision was performed 3 years after the initial presentation, followed by post-operative radiation due to positive margin. No evidence of disease recurrence was observed during the 3-year follow up.

Conclusion: Aggressive fibromatosis of head and neck is a challenging lesion due to its infiltrative nature, high propencity to recur and proximity to vital structures. Surgery should be well-planned using imaging and multidisciplinary approaches.

Keywords: Thailand, aggressive fibromatosis, desmoid, tracheal compression, mediastinum

J Med Assoc Thai 2018; 101 (6): 859-62
Website: http://www.jmatonline.com

Aggressive Fibromatosis (desmoid fibromatosis, or desmoid tumor) is a rare soft tissue tumor arises from aponeurotic or muscular structures. It is a benign but locally aggressive tumor with a high rate for local recurrence[1]. Allen classified aggressive fibromatosis as follows: extra-abdominal desmoids, abdominal wall desmoids, intra-abdominal desmoids, multiple desmoids and desmoids in Gardner's syndrome[2]. Enzinger and Weiss[1] classified fibromatosis into superficial and deep types. Superficial-type fibromatosis is found in palmar, plantar, penile and knuckle pad lesion. Deep-type fibromatosis is classified into the following 3 subtypes: extra-abdominal, abdominal and intra-abdominal. Head and neck fibromatosis are deep and extra-abdominal fibromatosis. The estimated incidence of fibromatosis in general population is two to four cases per million per year with approximately one-third of those being identified in extra-abdominal locations[3]. It was reported that approximately 12% of aggressive fibromatosis developed in the head and neck[4]. A literature review from 1968 to 2008 conducted by Kruse found only 179 published cases of head and neck fibromatosis, with a 30.04% recurrence rate among the 143 patients that were followed up[5]. The restricted and complex anatomy of the head and neck region and tumor proximity to head and neck vital structures contributed this tumor's aggressive clinical presentation, difficult management and high rate of recurrence. The present case is the first aggressive fibromatosis of neck and superior mediastinal area reported in Thailand.

Correspondence to:

Limviriyakul S. Department of Otorhinolaryngology, Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok 10700, Thailand.
Phone: +66-2-4198047, **Fax:** +66-2-4198044
Email: siriporn.lim@mahidol.ac.th

Case Report

A 76-year-old woman presented in 2005 with a large anterior neck mass and partial upper airway obstruction required tracheostomy. Tissue biopsy report was invasive follicular carcinoma of thyroid origin. The patients refused further treatment. In 2007 she developed dyspnea due to external compression of trachea below the tip of the tracheostomy tube, occluding 80% of the lumen. The existing tube was replaced with an extra-long tube and the patient refused further treatment. Three months later, the patient developed dyspnea from tracheal narrowing below the tube. An endotracheal tube was used to pass the narrowed area with the tip of the tube positioned 1 cm above the carina. She was then referred to the authors' tertiary referral hospital in 2008. The initial presentation at the authors' center, she had a 5 x 8 cm midline neck mass above the sternal notch,

How to cite this article: Chongkolwatana C, Limviriyakul S, Slisatkorn W, Pongsapich W. Aggressive fibromatosis of neck and superior mediastinum: A Case Report. J Med Assoc Thai 2018;101:859-62.

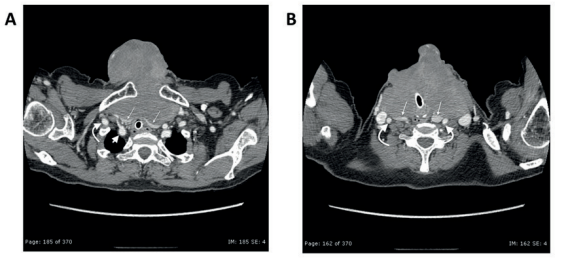

Figure 1. CT scan showed a large heterogeneous enhancing lobulated midline anterior neck mass protruding into subcutaneous fat tissue. The tumor also displaced both common carotid arteries (curved arrow) posteriorly and had pressure effect on thyroid gland (thin arrow). (short arrow-subclavian artery). (A) CT scan at superior mediastinal level; (B) CT scan at neck level.

with rubbery consistency and fixed to surrounding structures. She also had bilateral vocal folds paralysis. The original biopsy slides were reviewed by the institute pathologist, the revised and final diagnosis was aggressive fibromatosis.

CT scan showed a large lobulated heterogeneous mass at the anterior neck, that was displacing the thyroid gland and both carotid arteries (Figure 1). The inferior part of the mass extended to the mediastinum at the T-4 vertebral level. The mass showed no bony or vascular invasion except pressure effect on the trachea. Esophagography showed no evidence of esophageal invasion or obstruction. Mass excision and total thyroidectomy via cervical collar incision and median sternotomy were performed. The mass was inseparable from the thyroid gland and attached to bilateral common carotid arteries and internal jugular veins, thyroid cartilage, cricoid cartilage, trachea and brachiocephalic artery. The mass had markedly compressed the trachea without invasion. The mass was painstakingly dissected from surrounding structures, including the larynx, trachea, and vascular structures. Normal tissues located adjacent to the periphery of the tumor, including strap muscles were removed for clear margin whenever possible to enhance the likelihood of achieving a negative margin. Pathological analysis of the tumor revealed desmoid fibromatosis. Given that the posterior margin was found to be positive for tumor cells, the patient recieved postoperative radiation therapy (5000 cGy).

The patient was followed for 3 years with no recurrence. The tracheal collapse was recovered, but she still required regular-length tracheostomy due to bilateral vocal fold paralysis, for which she refused further treatment.

Ethical approval

The study was approved by the Institutional Review Board and Ethical Committee of the Faculty.

Discussion

Head and neck desmoids fibromatosis is a rare and aggressive disease. It is the subgroup of extra-abdominal fibromatosis, which only a few large series had been published [4, 6, 7]. A review of the literature published during 1968 to 2008 revealed only 179 published cases of head and neck fibromatosis[5]. Some studies reported no identifiable predilection for age group and gender, although the mean age seemed to be lower than the mean age of other types of fibromatosis [5, 6]. Other studied reported female predominance among head and neck cases [4, 8]. In the head and neck subset of fibromatosis, the most common location was reported to be the neck including the supraclavicular area, (Fasching, et al 71%, Conley et al 40%, Wang *et al.* 62%)[6-8], with the rest occurred at the scalp, face, oral cavity or sinuses[7] Patients usually presented with painless, non-ulcerative mass which grew steadily and sometimes rapidly[4]. Some patients may present with compressive symptoms of the surrounding structure such as pain, hoarseness or airway obstruction[4].

The etiology of Aggressive fibromatosis remains unknown despite many proposed causes including genetic[1], endocrine[9] and physical trauma, such as surgery and post-radiation. Desmoid tumors have been associated with antecedent trauma, particularly surgical interventions, in patients with familial adenomatous polyposis (Gardner syndrome)[1]. In the head and neck fibromatosis, trauma did not seem to be as high as abdominal tumors [4, 8]. Morphologically characterized, fibromatosis is a rubbery, grey mass with a poorly demarcated margin usually invades surrounding muscle. Some parts of the tumor may be covered with a thin fibrous wall with no true capsules. Histologically, fibromatosis consists of uniform spindle-shaped fibroblasts with a dense collagen matrix. There are no pleomorphisms or mitotic figures which help to differentiate it from fibrosarcoma.

Regarding diagnosis, fine-needle aspiration (FNA) can lead to the suggestion of fibromatosis but this method is limited due to many differential diagnoses for fibrous spindle cell tumor. Both CT and MRI showed well-defined soft tissue mass with no specific characteristics for fibromatosis, although MRI was better than CT scan showing distinct tumor margins. Either CT scan or MRI is usually required for treatment planning. Appropriate surgery can be planned after

the diagnosis is well established by core biopsy[10] or incisional biopsy[8] and permanent section. One report suggested surgical excision after imaging with definite diagnosis established from frozen section[7].

The mainstay of treatment for aggressive fibromatosis of head and neck is complete surgical excision with negative margin[7,8]. The rate of recurrence was lower in cases with complete excision than in those with incomplete excisions[7]. Desmoids tumors have no capsule and tend to infiltrate to deep structure which makes it difficult to identify the full extent of the tumor. Complete removal of head and neck fibromatosis is difficult, complicated and sometimes impossible due to its aggressive nature and its close proximity to vital structures in the head and neck area[8]. Recurrence seems to be higher in head and neck fibromatosis than in other parts of the body[11]. Radiation is indicated for inoperable tumor, gross residual tumor , positive margin and recurrent tumor with high risk reoperation[12]. Wang, et al reported high percentage of disease free patients having microscopical positive margin and complete spontaneous regression in some cases with gross residual tumor. As such, close follow up might be feasible and radiation may be reserved for tumor regrowth[8]. Age, gender and location were not found to be significant prognostic factors[5].

Even in the cases with pathologically-proven negative margin, recurrence is still possible. Close follow-up is recommended in all head and neck fibromatosis patients. Hoos, et al[10] suggested follow up examination every 3 months for 3 years, then every 6 months for 2 years and every year thereafter. CT or MRI should be used to investigate the difficult to examine locations and cases with suspected recurrence. Surgery, whenever possible, remains primary treatment for recurrent diseases. Despite delayed treatment for 3 years and positive margin by surgery, the present patient had no evidence of recurrence during the 3-year follow-up.

Conclusion

Aggressive fibromatosis of head and neck is a challenging lesion due to infiltrative nature, the high propensity to recur and the proximity to vital structures. Complete resection is still the mainstay of treatment. Surgery should be well-planned using imaging and multidisciplinary approaches. Despite the high rate of recurrence, the overall prognosis in aggressive fibromatosis of the head and neck is good.

What is already known on this topic?

Aggressive fibromatosis of head and neck is a rare disease. Complete surgical excision, which is the mainstay of treatment, is difficult and recurrence rate is high.

What is the study add?

Otolaryngologists should keep in mind that aggressive fibromatosis can present with an unusual local aggressive neck mass. Clinically, it can mimic the more common invasive thyroid cancer. Complete surgical removal, should be well-planned using imaging and multidisciplinary approaches.

Acknowledgement

The authors would like to thank Miss Jeerapa Kerdnoppakhun for manuscript preparation.

Potential conflicts of interest

The authors declare no conflicts of interest.

References

1. Enzinger FM, Weiss SW. Fibromatosis. In: Enzinger FM, Weiss SW, editors. Soft tissue tumors. Louis St.: Mosby; 1995: 201-29.
2. Allen PW. The fibromatoses: a clinicopathologic classification based on 140 cases. Am J Surg Pathol 1977; 1: 255-70.
3. Reitamo JJ, Hayry P, Nykyri E, Saxen E. The desmoid tumor. I. Incidence, sex-, age- and anatomical distribution in the Finnish population. Am J Clin Pathol 1982; 77: 665-73.
4. Masson JK, Soule EH. Desmoid tumors of the head and neck. Am J Surg 1966; 112: 615-22.
5. Kruse AL, Luebbers HT, Gratz KW, Obwegeser JA. Aggressive fibromatosis of the head and neck: a new classification based on a literature review over 40 years (1968-2008). Oral Maxillofac Surg 2010; 14: 227-32.
6. Conley J, Healey WV, Stout AP. Fibromatosis of the head and neck. Am J Surg 1966; 112: 609-14.
7. Fasching MC, Saleh J, Woods JE. Desmoid tumors of the head and neck. Am J Surg 1988; 156: 327-31.
8. Wang CP, Chang YL, Ko JY, Cheng CH, Yeh CF, Lou PJ. Desmoid tumor of the head and neck. Head Neck 2006; 28: 1008-13.
9. McDougall A, McGarrity G. Extra-abdominal desmoid tumours. J Bone Joint Surg Br 1979; 61-B: 373-7.

10. Hoos A, Lewis JJ, Urist MJ, Shaha AR, Hawkins WG, Shah JP, et al. Desmoid tumors of the head and neck - A clinical study of a rare entity. Head Neck 2000; 22: 814-21.
11. El-Sayed Y. Fibromatosis of the head and neck. J Laryngol Otol 1992; 106: 459-62.
12. Jelinek JA, Stelzer KJ, Conrad E, Bruckner J, Kliot M, Koh W, et al. The efficacy of radiotherapy as postoperative treatment for desmoid tumors. Int J Radiat Oncol Biol Phys 2001; 50: 121-5.