Case Report

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# Rare Undifferentiated Tumour of Thyroid: Primary Thyroid Fibrosarcoma

Surgery Section

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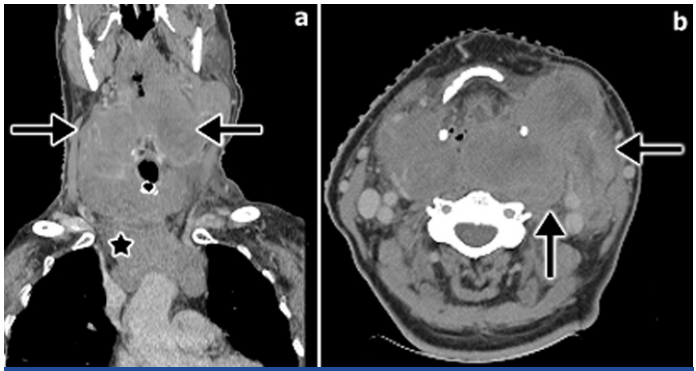
## ABSTRACT

Primary thyroid fibrosarcoma cases are very rare. Although it is a known fact that soft tissue sarcomas show slow growth, there have been some cases in literature similar to our case in which there was a fast-growing tumour tissue causing breathing and swallowing difficulties due to painless pressure. For diagnosis, there is no specific clinical or radiological finding. We report a 67-year-old male with a mobile fast-growing mass covering almost all over the neck that appeared 2 months prior to the admission. Laboratory findings showed that the patient was euthyroid. Fine needle aspiration biopsy results are consistent with suspicion of a mesenchymal, histiocytic, epithelial or lymphoid tissue origined malignancy. Patient was taken into surgical operation. The thyroid tissue invaded the main vascular structure, trachea and esophagus. Due to this situation R1 resection was applied. Immunohistopathological examination showed a conventional type of fibrosarcoma. After the surgery, radiotherapy and chemotherapy had been planned and applied. Patients died before the radiotherapy sessions ended. It should be kept in mind that a rapid growth in thyroid tissue can be thyroid fibrosarcoma, there could be a rapid clinical course and poor prognosis after operation.

**Keywords:** Euthyroid, Fine needle aspiration biopsy, Soft tissue sacrcoma

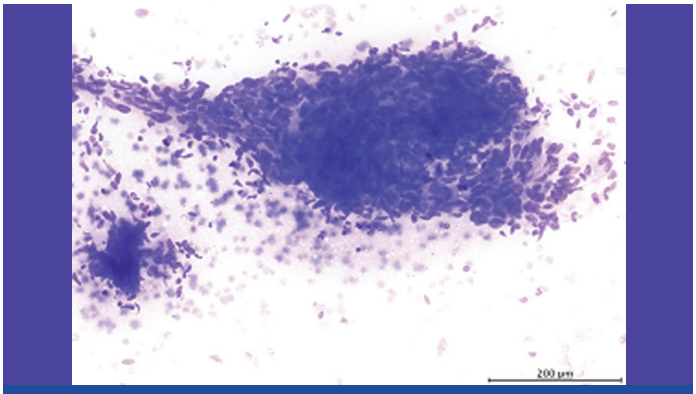
## CASE REPORT

A 67-year-old male was admitted to General Surgery Polyclinic of Dicle University Hospitals with a mobile fast-growing mass that appeared 2 months prior to admission. He also had apparent respiratory distress and dysphagia. Clinical examination showed a solid, mobile mass presented almost all over the neck extending beneath the clavicle and palpated easily. Ultrasound examination revealed heterogeneous looking masses with irregular borders, which were extending to left mandibular area. These masses were larger than 4cm in diameter. Heterogeneous, hypodense mass appearance was detected at thyroid parenchyma, which had calcific foci and was deviating trachea to the right while surrounding and narrowing at anterior region via neck-computed tomography. The mass had a size of 54x48mm at right and was extending to retrosternal area with a size of 69x58mm at left [Table/Fig-1a,b]. Complete Blood Count (CBC), thyroid function tests and related biochemical tests including serum glucose, urea, creatine, AST, ALT, ALP, $Ca^{2+}$ and arterial blood gas were all in normal range. Fine Needle Aspiration Biopsy (FNAB) results revealed atypical cells with a fusiform-epithelioid shaped anucleolate irregular nucleus, lacking cytoplasm [Table/Fig-2].

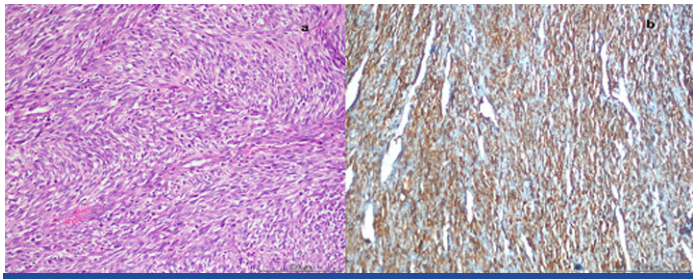



**[Table/Fig-1]:** Axial and coronal CT sections of thyroid gland. Infiltrate hypodense mass monitored in anteior, posterior and lateral regions of both thyroid glands. (a) coronal. Hypodense mass on the left presses on the trachea (arrows). At the upper media stinum conglomerated lymphadenopathies (star). (b) Axial. Lesion surrounded the trachea at the anterior (arrows).

This led us to suspect a mesenchymal, histiocytic, epithelial or lymphoid tissue origined malignancy. Surgical intervention was an obligation as the patient had respiratory distress and dysphagia. During the surgical procedure, it was observed that substernal located thyroid tissue was large in size (420g) and gray in colour with moderate hardness. It invaded the main vascular structure, trachea and esophagus. Due to this situation, R1 resection was applied without damaging recurrent nerves. There was no major complication after surgical procedure. Immunohistopathological examination showed a conventional type of fibrosarcoma, staining negative with EMA, LMWCK, PanCK, calcitonin, CEA, CD31, NSE, SMA, HMB45 dye and positive with vimentin [Table/Fig-3a,b]. At




**[Table/Fig-2]:** Spindle-shaped atypical cells at fine niddel aspiration (giemsa, 200X).




**[Table/Fig-3]:** a) Spindle-shaped atypical cells arranged facicules (200X, H&E); b) vimentin positivity in tumour cells (immonohistochemistry, 200X).

the beginning 2 Gy/frc total 50 GY radiotherapy (RT) to region and cervical lymph nodes with tomotherapy, and then ifosfamide 2500 mg/m$^2$, mesna: 2500 mg/m$^2$, doxorubicin 60 mg/m$^2$ treatment for a period of 21 days once a day was given.

During 18 session of radiotherapy, dyspnea occurred due to tracheal oedema and hence tracheostomy was opened. In CT, it was observed that thyroid region was still filled with necrotic tumour tissue. Patient had 3 more RT sessions after leaving intensive care. Then, the general condition of the patient worsened at the 21$^{st}$ session of radiotherapy, and was taken to intensive care immediately, died due to cardiopulmonary arrest.

## DISCUSSION

Primary thyroid fibrosarcoma cases are very rare. Thyroid sarcomas constitute less than 1% of all thyroid tumours [1,2]. This situation complicates both diagnosis and treatment. Fibrosarcoma can be seen over the age of 40, regardless of gender, making a peak during the ages 60-79 [3,4] with an exception of a case reported by Postovsky et al., a 14-year-old paediatric patient with undifferentiated thyroid sarcoma [5]. In this case, the age of the patient was in accordance with the literature. Surov et al., observed that thyroid fibrosarcoma constitutes 9.2% of all thyroid sarcomas [3]. For diagnosis, there is no specific clinical or radiological finding. Although it is a known fact that soft tissue sarcomas show slow growth, there have been some cases in literature similar to our case in which there was a fast-growing tumour tissue leading to breathing and swallowing difficulties due to painless pressure [4,6]. Darouassi et al., reported a case with a fast growing mass in thyroid tissue covering entire neck within 2 months prior to admission to the hospital. Similar to this report, present case also had a fast growing mass covering entire neck within 2 months [6]. General complaints were mass in the neck, difficulty in swallowing, shortness of breath, and our case was similar to the other case's presentations [1,4,6]. Euthyroid laboratory findings of our case were also similar to other cases in literature [1,4,6].

Although US and CT have the ability to show the mass and give us an idea about its size, they are not specific while supporting the diagnosis. Surov et al., reported that sarcomas can be seen hypo- or hyperechoic in US and hypo or hyperdense in CT [3]. In the same study, they also reported that both US and CT findings can reveal the growth in the entire thyroid tissue as well as its pressure on trachea and esophagus more accurately. Atypical cells were identified by FNAB. The most important finding for pre-operative diagnosis was that these cells could be of mesenchymal, histiocytic, epithelial or lymphoid origin.

With FNAB, atypical cells were identified, and these pre-operative findings showed that these cells could be mesenchymal, histiocytic, epithelial or lymphoid originated. Titi S et al., performed FNAB and suspected of schwannoma, which was one of the powerful findings for indicating a mesenchymal origin [1]. Nevertheless, all of these findings are not enough for definitive diagnosis. Definitive diagnosis of fibrosarcomas should be made with immunohistochemical methods [7]. Prognosis is closely related with the structure of tumour cells, cellular pleomorphism, mitotic activity and necrosis [1]. Cells with minimal atypia and view of spindle cell neoplasm containing lots of collagen fibers are remarkable in Hematoxylin and eosin staining. In immunohistochemical staining, these spindle cell areas were stained positively with vimentin, and this also supports our diagnosis [1,4,6,7]. Furthermore, negative staining with EMA is another clue that supports the differential diagnosis of carcinoma and conventional type fibrosarcoma. Besides, negative staining with HMB45 plays an important role for the differential diagnosis of clear cell carcinoma and malignant melanoma [1].

Chemotherapy and radiotherapy has been employed especially in subset of cases that cannot be totally resected, but their benefit is controversial. In addition to these situations during RT treatment, the mass resumed to grow and fill thyroid region rapidly. As in this case, patients generally die within 6 months after surgical intervention [4,6,8].

## CONCLUSION

Primary thyroid fibrosarcomas are very rare among thyroid malignancies. Therefore, there is no consensus on diagnosis, clinical course and prognosis. In this regard, we find the sharing of clinical experience very valuable. It should be kept in mind that a rapid growth in thyroid tissue can be thyroid fibrosarcoma and unlike other soft tissue sarcomas, there could be a rapid clinical course and poor prognosis after operation.

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