Aggressive Fibromatosis of the Neck.

Case Presentation.

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OUTLINE.

• The clinical picture of head and neck fibromatosis.

• Etiology.

• Pathology.

• Current opinion regarding its treatment.
Case report.

- A 14-year-old boy who had a 1 year history of a painless mass overlying his right neck.
- Referred as a case of Desmoid tumor (Incisional biopsy-histology) from Tamale Hospital April 2010.
- The patient had no prior medical or surgical history.
- Family Hx not significant.
- A history of remote minor neck trauma was present.
- No known allergies.
- No history of regular Rx.
- No dysphagia, no cough, no fever, no wt. loss, no night sweats.
Physical examination,

- General condition – satisfactory, afebrile, not pale, anicteric, not wasted, well hydrated.
- There was a smooth firm, non-tender mass overlying his RT neck, extending:
  lateral border- posterior triangle Rt, 
  medial-anterior triangle of the contralateral side, 
  superior-level II neck Rt, 
  inferior- clavicular region Rt, fixed to underlying structures, there was no plane b/n the tumor and clavicular bone on palpation.
- Post incisional biopsy scar about 4 cm long noted over the RT side neck.
- The laryngo-tracheal framework was shifted to Lt. side of the neck.
- No dyspnoea, RR 18/min
- No hoarseness, IDL – anatomical position of the glottic structures displaced, TVC not visualized.
- No palpable lymph nodes.
• Other systemic examination –

• *EENT, CVS, Chest, Abdomen-* NAD

• *CN I-XII* intact.

• Neurologic symptoms, mainly tingling in the RT arm, had been present for a few months and were most likely related to involvement of the brachial plexus.

• There was no evidence of any other primary malignant tumor.
The differential diagnosis:

- Rhabdomyosarcoma
- Fibrosarcoma
- Lymphoma
- Metastatic disease.
Patient’s work-up.

- **FBC**-Hb, WBC (T.,D.,) INR, BF comment - NAD
- **BUE** - NAD
- **LFT**- NAD
- **X-Ray soft tissue NECK** (AP, LAT)- Radio- Opaque mass RT neck, Laryngo-tracheal framework shifted to contralateral side significantly.
- **X-RAY Chest** (AP, LAT)- NAD
- **USG Neck** (Soft tissue solid mass, compressing great vessels of the neck on the ipsilateral side, suggestive of Lymphoma)
- **Incisional biopsy(re-biopsy)**( proliferation of benign fibroblast type cells with edematous stroma and small areas of chronic inflammation. No evidence of malignancy. Consistent with fibromatosis.) -02.06.10
- **CT scan** head, neck, chest, abdomen.
- **Panendoscopy**.
Pathologically, the tumor demonstrated a moderately cellular mixture of myofibroblasts and fibroblasts arranged in long sweeping bundles, within an abundant collagen matrix.
No areas of necrosis or pleomorphism were seen and there were no mitotic figures. The diagnosis of aggressive fibromatosis was made.
• Two unusual features of this case were the site of the lesion and its rapid growth without airway obstruction.
Patient was presented at Head and Neck Tumor Board KATH.

- **Decision:** Patient was considered as a poor surgical candidate, the following alternative modality of treatment was considered:
  - Low-morbidity therapy aimed at inhibiting the growth of this unique tumor.
  - Initial treatment to start with: Chemotherapy +(-) RT.
  - For complete excision after assessment post treatment with above Rx and CT scans.
Patient’s treatment.

• Currently patient is under care of Oncology Department with following treatment:

• Low dose Chemotherapy:
  weekly low-dose i.v. methotrexate and vinblastine for 12 months.

• Close monitoring for myelotoxicity, hepatotoxicity, UAO.

• To review of the patient at H&N Tumor Board after completion of the treatment.
Aggressive fibromatosis

Also known as extra-abdominal desmoid is part of the spectrum of fibrous tissue proliferative disorders.
There is a progression in the microscopic picture of fibrous tissue proliferations.

- The normal reparative scar formation
- Fibroma
- Keloid
- Pseudosarcomatous fasciitis
- Fibromatosis
- Differentiated fibrosarcoma
- Undifferentiated fibrosarcoma
• Aggressive fibromatosis occupies the middle position of that spectrum, and is distinguished from fibrosarcoma by its

• **lack of anaplastic cells and absence of distant metastasis.**
The World Health Organization (WHO) classifies Desmoid tumours as somewhere in between truly benign tumours and the malignant tumours.
Aggressive fibromatosis represents a locally infiltrative process composed of mature fibroblasts and collagen, without evidence of cellular anaplasia or abnormal mitotic figures.
Desmoid fibromatosis may occur as **Abdominal**
**Intra-abdominal and**
**Extra-abdominal locations.**

- Extra-abdominal fibromatosis most frequently occurs in the limbs, followed by the head and neck region.

- Fibromatosis is a rare entity, representing only **0.03%** of all neoplasms.

- Extra-abdominal desmoids represent about 1/3 of all desmoid tumors

- About 3 to 15% arise in the head and neck.
What causes aggressive fibromatosis?

• **Still under investigation.**
• Mutation in the *APC* gene
• Inherited (as several family members may be diagnosed with them.)
• In surgical scars after surgery.
• Pregnancy (estrogen/hormonal changes).
• M:F
• Any age
• No predilection for any ethnic group.
The management of desmoid tumors

- Desmoid tumours are very individual in their responses to treatment, and no one treatment alone seems to have a high success rate.
- Surgery (S)
- Radiation therapy (RT)
- Cytotoxic and noncytotoxic chemotherapy.
- S+RT
- S+ChemoRx
- RT+ChemoRx
- S+RT+ChemoRx
Total resection with wide margins is reportedly the best treatment.

- However, the anatomy of the head and neck makes such resection difficult.

- Incomplete resection is known to result in higher tumor recurrence than complete resection.

- The recurrence or progression of a tumor in the H&N region is known to cause mortality by compression of the airway or major blood vessels.
Surgical treatment strategies aim:

• Achieving local control while preserving proper function and providing with an adequate cosmetic result.

• When medically and technically feasible, complete resection of the tumor with negative microscopic margins is the first-line treatment it must be noted that the importance of positive margin status to local recurrence rates.
Radiotherapy.

• For patients with inoperable tumours, or those where surgery may result in major functional loss, radiotherapy is a reasonable option.

• Improved disease control has been reported in both adjuvant and primary settings.

• Primary radiotherapy has achieved a local control rate of up to 93%.
Systemic therapy includes:

- **Noncytotoxic**:
  - Tamoxifen
  - Testolactone
  - Nonsteroidal anti-inflammatory drugs

- **Cytotoxic**:
  - Methotrexate
  - Vinblastine
  - Doxorubicin
CONCLUSIONS:

• Attempts to achieve negative resection margins may result in unnecessary morbidity and may not prevent local recurrence.

• Operations that preserve function and structure should be the primary goal, because the presence of residual disease cannot be clearly shown to impact adversely on 5-year disease free or overall survival.
In summary

- Desmoid tumors (aggressive fibromatosis) are benign neoplasms with high rates of recurrence after surgery.
- Complete surgical resection followed by radiation therapy is the current standard therapy.
In summary (cont.)

• Treatment strategies must be individualized so as to provide adequate local control with subsequent acceptable functional and esthetic outcome.

• Despite the applied therapeutic method, due to the high recurrence rate, these patients must be closely followed by clinical examination and radiographic studies.
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Competing interests.

• The authors declare that they have no competing interests.
Thank you!
Rx with Chemotherapy

FIG 1. Computed tomographic scan before chemotherapy showing a huge tumour compressing the airway, vessels, and oesophagus after debulking surgery and tracheostomy.

FIG 2. Computed tomographic scan after 16 months of chemotherapy showing excellent tumour shrinkage and removal of the tracheostomy tube.