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Case Report

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Sudden Onset Breathlessness in a young boy:An Uncommon Aetiology

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ABSTRACT

Schwannomas are the most common mediastinal neurogenictumors; they are usually benign and slow growing and frequentlyarise from a spinal nerve root, but may involve any thoracicnerve

Introduction:

Aschwannoma,also known as neurilemoma or neuroma is a benign nerve sheath tumour composed of schwanncells,whichnormally produce the insulating myelin sheath covering peripheral nerves.

Case Report:

An eleven year old boy required urgent admission in our department on 13/03/2012 with complaints of progressive shortness of breath for last two months along with left sided chest discomfort and dull ache for same duration and increasing swelling of left anterior chest for last one month.On examination he was dyspnoeic(MMRC-Grade 3),Respiratory rate 36/min, Pulse-120/min, BP-108/60 mmofHg.There was no clubbing,nolymphadenopathy.There was diffuse bulging of left anterior chest and a median sternotomyscar,prominentepigastricpulsation.Trachea was on right side,dull left hemithorax with decreased breath sounds.An Ultrasonography of thorax prior to attempt at thoracic therapeutic thoracocentesis showed a large mass in anterior mediastinum(16x13x13.5 cm),Bilateral pleural effusion(small) and minimum pericardial effusion.On **past history** the child required several hospitals admissions prior to this.Eight months back(27/05/11) he was first admitted in this hospital with similar complaints and a retrosternal chest pain,unproductivecough,low grade fever for three months.At that time screening Echo revealed moderate pericardial effusion,increasing**tamponade** from which guided aspiration was done(300 ml straw coloured fluid).On analysis of the pericardial fluid,the cell count was 1900/cmm(Neutrophils 10%, Lymphocytes-85% and others 5%),Sugar 45mg/dl,protein 4.4 gm/dl and ADA 80.7 U/L,PAP stain was negative for malignancy.He was advised ATD: CAT I(DOTS)+prednisolone.There was symptomatic improvement of breathlessness and he was discharged with advice for follow up at cardiac & chest OPD of this institute.About two months later(14/07/2011) he was again brought in this hospital with increased sudden onset breathlessness.A repeat ECHO was done(15/07/2011) which showed an extra cardiac mass in anterior mediastinum causing external compression on right ventricular outflowtract.FNAC from mediastinal SOL(04/08/11) was suggestive of **Teratoma**.So biopsy was suggested.Among all biochemical parameters serum Alpha Feto protein level was 1.38 ng/ml Serum hcG + β subunit level being < 0.60 mIU/ml (< 2.6).

Next step was surgery.The anterior mediastinal mass was exised surgically on 17/08/11.

The boy made an uneventful recovery and was discharged after 39 days.Now in the current scenario as there was rapid re-growth of the otherwise benign schwannoma question arises wheather there was any malignant degeneration of it or not.So the opinion of tumour board was also required.On 30/03/2012 The Tumour Board suggested Review of the HP slides,Repeat biopsy/Re exploration to get sufficient material for HPE and to relieve symptom.InjEndoxan(700 mg)I.V single dose was advised.

Discussion:

Schwannomas are homogenous tumours,consisting only of schwann cells. The tumour cells always stay on the outside of the nerve,but the tumour itself may either push the nerve aside and or up against a bony structure(therby possibly causing damage).Schwannomas are relatively slow growing.For reasons not yet understood,schwannomas are mostly benign and less than 1% become malignant,degenerating into a form of cancer known as neurofibrosarcoma.Schwannomas can arise from a genetic disorder known as neurofibromatosis.Theyare universally S-100 positive.S-100 proteins are a family of low molecular weight proteins found in vertebrates and characterized by two calcium binding sites that have ‘’helix-loop-helix’’(EF-hand type) conformation.They are encoded by a family of genes whose symbols use the S100 suffix,for example S100A1, S100A2,S100A3.They are also considered as Damage-associated molecular pattern molecules(DAMPs).Most S100 proteins are homodimeric,consisting of two identical polypeptides,which are held together by non covalent bonds.S100 proteins are structurally related to calmodulin.On the other hand they differ from calmodulin on the other features.Forinstance,their expression is cell specific,i.e they are expressed in particular cell types.Their expression dependes on environmental factors.Tocontrast,calmodulin is a ubiquitous and universal intracellular Ca2+ receptor widely expressed in other cells.Schwannomas can be removed surgically.Recurrences after total removal are rare.Verocay(Antoni A’’ bodies are seen histologically in schwannomas.They are cellular areas surrounded by nuclear palisades(as opposed to less cellular ‘’Antoni B’’.

References:

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Source:Department of Medicine, Veterans General Hospital-Kaohsiung, Taiwan, R.O.C

2.Benign giant mediastinalschwannoma presenting as cardiac tamponade in a woman: a case report

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