

may also have to be considered, including paraganglioma, branchial cleft cyst, malignant lymphoma, metastatic cervical lymphadenopathy etc³. Tumors of the parapharyngeal space are rare, with neurogenic tumors being the most common. Schwannomas account for 55% of these tumors, approximately half of which arise from the vagus nerve⁴. Neoplasms of the vagus nerve include paragangliomas (50%), schwannomas (31%), neurofibromas (14%), and neurofibrosarcomas (6%)⁵.

Occasionally, a paroxysmal cough may be produced on palpating the mass. This unique clinical sign, associated with a mass located along the medial border of the sternocleidomastoid muscle, should make clinicians suspicious of vagal nerve sheath tumours^{1,6}.

The usefulness of FNAC is still controversial; the majority of authors do not recommend open or needle biopsy for these masses³. In our case, a FNAC was performed but was inconclusive.

Imaging can be very useful in differentiating vagal schwannomas from other lesions in this area, allowing the surgeon to plan the operative procedure to remove these tumors. On CT images, vagal schwannomas appear as well-defined masses, usually of higher attenuation than muscle on contrast-enhanced images².

Histologically, two main patterns, Antoni-A (Verocay body formation) and Antoni-B (no distinctive cell or fibre pattern) have been described. A mixed picture of both types can exist².

Treatment of vagal nerve tumours is complete surgical excision with preservation of the neural pathway. Incomplete treatment, such as open biopsy, should be avoided, since it makes definitive excision of the tumour much more difficult¹.

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Bardet-Biedl Syndrome: A rare Case Report from Rural Medical College.

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Abstract : Bardet-Biedl syndrome (BBS) is a genetically heterogeneous autosomal recessive disorder characterized by progressive retinal dystrophy, polydactyly, obesity, hypogonadism, mental retardation, and renal dysfunction. We present a rare classical case with characteristic features of four major and four minor criteria along with a brief review of the literature.

INTRODUCTION

The Bardet-Biedl syndrome (BBS) is a rare ciliopathic human autosomal-recessive disorder. Its incidence 1:1,60,000. A classical case of BBS features such as marked central obesity, retinal dystrophy, polydactyly, overcrowded teeth, high arched palate, VSD and mental retardation a with a brief review of the literature, is presented.

CASE REPORT

24 year old female with central obesity presented to our department for USG abdominopelvis study with a history of fever, dimness of vision and irregular cycles. USG revealed hepatosplenomegaly with ventricular-septal defect. On examination patient had central obesity, oscillating nystagmus, post-axial polydactyly in both hands and feet, overcrowding of teeth, high arched palate and mild mental retardation. On ophthalmic examination b/l consecutive ascending optic atrophy with retinitis pigmentosa. therefore, the diagnosis of Bardet-Biedl syndrome was made.

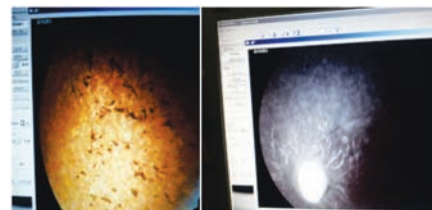
DISCUSSION

Bardet-Biedl syndrome (BBS) was initially described by Bardet and Biedl(1920). Laurence and Moon (1865) syndrome is a separate entity because the patients of Laurence and Moon had paraplegia, but no polydactyly and obesity.

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Postaxial polydactyly with hexadactyly of hands and feet



Fundus photography with features of retinitis pigmentosa



High arched palate