New criteria for improved diagnosis of Bardet-Biedl syndrome: results of a population survey. J Med Genet. 1999;36: 437-46.
5. McLoughlin TG, Krovetz LJ, Schiebler GL. Heart disease in the Laurence-Moon- Biedl-Bardet syndrome: A review and a report of three brothers. J Pediatr. 1964;65:388-99.
6. Ross AJ, Beales PL. Bardet-Biedl syndrome [internet] Gene Reviews. Available from *http://www.geneclinics.org/profiles/all.html.* [cited 2008 December]. Accessed on 1 January, 2013.
7. Yang Z, Yang Y, Zhao P, Chen K, Chen B, Lin Y, *et al*. A novel mutation in BBS7 gene causes Bardet-Biedl syndrome in a Chinese family. Mol Vis. 2008;14:2304-8.

Meningitis due to *Neisseria meningitidis* Serogroup B in India

MEENAKSHI AGGARWAL, VIKAS MANCHANDA AND *B TALUKDAR

*From the Departments of Clinical Microbiology and Infectious Diseases Division, *Pediatric Medicine, Chacha Nehru Bal Chikitsalaya, Raja Ram Kohli Marg, Geeta Colony, Delhi, India.*

Correspondence to: Dr Vikas Manchanda, Room # 101, Clinical Microbiology and Infectious Diseases Division, Chacha Nehru Bal Chikitsalaya, Raja Ram Kohli Marg, Geeta Colony, Delhi 110 031, India. microcnbc@gmail.com
Received: September 29, 2012; Initial review: October 22, 2012; Accepted: January 24, 2013.

Invasive meningococcal disease has a fulminant course and high mortality. *Neisseria meningitidis* serogroup A is predominantly responsible for meningococcal disease in India and the developing countries. Group B meningococcus, which is prevalent in the developing world is uncommon in India. We herein report the second case of group B meningococcal infection from the country, two decades after the reporting of the first case. Ineffective vaccines against serogroup B warrant the need for close surveillance of this disease.

Key words: *Child, India, N. meningitides serogroup, Surveillance.*

Invasive meningococcal disease commonly follows a fulminant course and has high mortality [1]. Thirteen serogroups of *Neisseria meningitidis* have been identified, but six of these serogroups (A, B, C, W135, X and Y) are responsible for majority of the infections worldwide [1]. Serogroup A strains are predominantly responsible for meningococcal disease in developing countries, including India [2]. Serogroup B strains are responsible for outbreaks of meningitis in the developed world where vaccines against serotypes A,C,Y and W135 are extensively used [3]. Group B meningococcus is not prevalent in India, with only one previous report [4]. We herein report the second case of group B *N. meningitidis* infection from the country.

CASE REPORT

The patient was a one-year-old, boy weighing 7kg who presented to the pediatric emergency with seizures, history of high-grade fever, vomiting, lethargy and decreased oral acceptance since three days. He had multiple episodes of generalized tonic clonic seizures in last 24 hours. He was delivered at full term through an uneventful vaginal delivery. Immunization history was appropriate for age. No history of similar illness was present in the family and immediate contacts. On examination, child was conscious, had no cyanosis and had bilaterally constricted pupils with sluggish reaction to light. He was febrile (101^0F) with heart rate of 172 beats per minute and respiratory rate 42 per min. Capillary filling time was less than 3 sec. Anterior fontanelle was full and pulsatile. Neck rigidity was present. There was increased tone in all four limbs, deep tendon reflexes were brisk with bilateral extensor plantars. He had no skin rash. Initial clinical diagnosis of meningitis was made and therapy with intravenous ceftriaxone and anticonvulsants was started, in addition to supportive management.

Laboratory reports revealed that the child had hemoglobin of 8.1 g/dL with total white blood cell count of 10,610/mm^3 (56% neutrophils, 38% lymphocytes, 3.9% monocytes and 1.2 % eosinophils), and platelet count of 5.9 lakh/mm^3; C-reactive proteins was raised (178.97 mg/L). The blood pro-calcitonin levels were 118.23 ng/mL (≥10 ng/mL and plasma lactate levels were also raised (30.5 mg/dL). Renal function tests and serum electrolytes were within the normal range. The cerebrospinal fluid showed raised protein levels (113 mg/dL), and low levels of glucose (26 mg/dL). CSF cytology could not be reported because of hemmorhagic nature of tap. CSF lactate levels were increased at 83.93mg/dL and CSF chloride levels were 123 nmol/L. Latex agglutination was performed on the CSF sample and was

reactive for *N. meningitidis* group B (Pastorex Meningitis, *BIO-RAD*). Blood and CSF culture grew *N. meningitidis* as identified by Vitek 2 Compact system (BioMerieux, France). Serogrouping was done by *N.meningitidis* antisera (Remel Europe Ltd. UK) and was confirmed was as *N.meningitidis* serogroup B. The strain was found resistant to penicillin (MIC, 0.5 µg/mL) and ciprofloxacin (MIC, 0.5 µg/mL), and sensitive to ceftriaxone (MIC, 0.094 µg/mL), chloramphenicol (MIC, 0.19 µg/mL), azithromycin (MIC, 0.5 µg/mL), rifampicin ((MIC, 0.032 µg/mL), and meropenem (MIC, 0.032 µg/mL) as determined using E test and interpreted in accordance with CLSI guidelines. Blood and CSF samples tested positive for *ctrA* gene for *N.meningitidis* by Real time PCR assay. DNA extraction from blood and CSF samples was performed using the Magnapure Compact automated nucleic acid extraction system. (Roche Diagnostics, Basel, Switzerland) as per manufacturer's protocol. A 111 bp region of *ctrA* gene was amplified using ctrA specific primers, with slight modification [6]. *N. meningitidis* ATCC 13090 was used as the positive control.

The cranial ultrasonography showed slight ventricular prominence with normal cerebral parenchyma. The patient was placed under isolation and chemoprophylaxis with ciprofloxacin given to the close contacts of the patient. The patient was continued on intravenous ceftriaxone and phenytoin. He was put on mechanical ventilation because of repeated seizures and declining oxygen saturation. High grade fever and seizures were persisting till the 5th day of admission despite midazolam infusion (2µg/kg/min). Intravenous dexamethasone 1 mg 8 hourly was initiated along with other therapy on day 5. The patient started showing clinical improvement from the 7th day onwards with no fresh seizures, repeat blood culture showing no growth, and improved blood counts. Midazolam was discontinued on 9th day of admission. After 16 days of antibiotic therapy, repeat CSF examination was within normal limits and culture did not yield any growth. Despite the clinical and laboratory improvement, the antibiotic therapy was maintained for a total of 21 days. Dexamethasone was discontinued after 10 days of therapy. By day 29, the child had recoverd and was discharged on oral anticonvulsants from the hospital. Meningococcal carriage screening of the patient and the parents did not yield any positive results.

DISCUSSION

Meningococcal meningitis is a serious infection and if untreated, may be fatal with case fatality rates reaching 5-10% in developed countries and upto 20% in developing countries [7]. The neonate in the previous study had died within 6 hours of admission. However, in our study, the child survived and recovered after a prolonged hospital stay. The source of Group B meningococcus could not be ascertained in this case. The reason for very low prevalence of serogroup B in India is not known. It may be postulated that predominance of serogroup A might lead to suppression of serogroup B in the Indian population similar to the phenomenon observed in pre-vaccine era in developed countries. In addition to the varying geographical distribution, the proportion of cases caused by each serogroup may also vary by age. US studies describe serogroup B to be causing 30-50% of cases in infants younger than 1 year of age, while serogroups C, Y, and W135 causing 75% of meningococcal disease in those 11 years and older [7,8]. Both the cases from India (including the present case) were below one year of age.

Effective vaccines are available for meningococcal serogroups A, C ,Y and W135. Consequently, serogroup B, *N. meningitis* has become the major cause of bacterial meningitis especially in countries where vaccine for other serotypes has been introduced [4]. Vaccine against serogroup B strains for global use has been challenge. This is due to frequent antigenic variations among this serogroup. Antigenic mimicry of serogroup B polysaccharide with human neurologic tissues is also a problem [9]. Vaccines based on other bacterial cell components have shown poor protective immune response in children under 24 months of age [10]. Thus, the occurrence of even a single case Group B meningococcal meningitis in India has important public health implications.

Indian data on meningococcal disease is sparse and is limited to the studies undertaken during or immediately after suspected outbreaks. Studies during inter-epidemic period and constant surveillance of the invasive meningococcal disease can help understanding the epidemiology of this highly fatal disease. Though there is a gap of two decades between the first and the second case report, there is a strong need for close surveillance and documentation for further group B meningococcal infections in India. With recent licensing of conjugated quadrivalent vaccines in the country, it will be interesting to observe changing epidemiology of invasive meningococcal disease in India.

Contributors: All the authors have prepared, designed and approved the study.
Funding: None; *Competing interests*: None stated.

REFERENCES

1. Manchanda V, Gupta S, Bhalla P. Meningococcal disease:

History, epidemiology, pathogenesis, clinical manifestations, diagnosis, antimicrobial susceptibility and prevention. Indian J Med Microbiol. 2006;24:7-19.
2. Sinclair D, Preziosi MP, John TJ, Greenwood B. The epidemiology of meningococcal disease in India. Trop Med Int Health. 2010;15:1421–35.
3. Khatami A, Pollard AJ. The epidemiology of meningococcal disease and the impact of vaccines. Expert Rev Vaccines. 2010;9:285–98.
4. Suri M, Kabra M, Singh S, Rattan A, Verma IC. Group B meningococcal meningitis in India. Scand J Infect Dis. 1994;26:771-3.
5. Corless CE, Guiver M, Borrow R, Edwards-Jones V, Fox AJ, Kaczmarski EB. Simultaneous detection of *Neisseria meningitidis, Haemophilus influenzae,* and *Streptococcus pneumoniae* in suspected cases of meningitis and septicemia using real-time PCR. J Clin Microbiol. 2001;39:1553–8.
6. Initiative for Vaccine Research. Bacterial Infections. Meningococcal Disease. Available from: URL: http://www.who.int/vaccine_research/diseases/soa_bacterial/en/index1.html. Accessed on September 1, 2012.
7. Rosenstein NE, Perkins BA, Stephens DS, Lefkowitz L, Cartter ML, Danila R, *et al*. The changing epidemiology of meningococcal disease in the United States, 1992–1996. J Infect Dis. 1999;180:1894-901.
8. CDC. Prevention and Control of Meningococcal Disease: Recommendations of the Advisory Committee on Immunization Practices (ACIP). MMWR 2005;54 (No. RR-7).
9. Granoff DM. Review of meningococcal group B vaccines. Clin Infect Dis. 2010;50:S54–65.
10. Tappero JW, Lagos R, Ballesteros AM, Plikaytis B, Williams D, Dykes J, *et al*. Immunogenicity of two serogroup B outer-membrane protein meningococcal vaccines: a randomized controlled trial in Chile. JAMA. 1999;281:1520-7.

Myxoid Lipoblastoma

JANANI KRISHNAN, VARUN HATHIRAMANI, *MEENAL HASTAK AND RAJEEV G REDKAR
*From the Department of Pediatric Surgery and *Histopathology, Lilavati Hospital and Research Centre, Mumbai, India.*
jankrish.doc@gmail.com

Correspondence to:
Dr Rajeev G Redkar, Consultant Pediatric Surgeon,
Lilavati Hospital and Research Centre,
Mumbai, India. rajeev.redkar@gmail.com
Received: December 10, 2012;
Initial review: December 21, 2012;
Accepted: January 24, 2013.

A rapidly growing soft tissue mass in the axilla of an infant raises the suspicion of a lipoblastoma or a liposarcoma. Excisional/incisional biopsy is vital in confirming the diagnosis and hence avoiding aggressive extirpation. This case report highlights the role of histopathology and immunohistochemistry as the gold standard in differentiating a lipoblastoma from a liposarcoma. In some cases where the histopathology is inconclusive, genetic rearrangement of the PLAG1 (pleomorphic adenoma gene 1) oncogene on chromosome 8q12 helps in confirming the diagnosis of lipoblastoma.

Key words: *Axillary mass, Lipoblastoma, Infant.*

The presence of an axillary mass in infancy entertains the diagnosis of a cystic hygroma, hamartoma or a soft tissue neoplasm. Lipomatous tumors, namely lipoblastomas are rare, benign tumors of infancy and early childhood. They arise from the embryonal fat cells which persist and continue to proliferate into postnatal life. They are characterized by their rate of rapid growth, local invasion and increased incidence of local recurrence of 14-25% [1]. The diagnosis of a liposarcoma in infancy should be made with caution, owing to its rarity and the aggressive treatment involved. The role of excisional biopsy, histopathology with immunohistochemistry and cytogenetics in the establishment of an accurate diagnosis, is highlighted in the following case report.

CASE REPORT

An 8-month-old female child presented to us with a swelling in the right anterior chest wall, extending through the axilla to the back (***Fig.* 1**). It was noticed since the age of 5 months, with rapid increase in size over the last one month, to the present size. The mass was non tender, soft to firm in consistency, bosselated surface, with no skin changes. The differential diagnoses included a vascular hamartoma, cystic hygroma, a soft tissue tumor such as lipoblastoma or a liposarcoma, or a matted lymph node mass.

The ultrasound examination showed an 8×7.5×4.5cm iso- to hyperechoic, lobulated solid mass, with posterior border extending beneath the lower margin of the scapula. MRI chest confirmed an 8×5×4cm right shoulder girdle,