

was seen in the epigastrium at the port site. Tenderness, warmth and erythema were present over the swelling. However there was no fluctuation. An ultrasound abdomen was done which showed anterior abdominal wall abscess with an approximate volume of 120 ml with no intra peritoneal extension was noted. CECT abdomen was done to confirm that the abscess cavity was not communicating with peritoneal cavity. Ultrasound guided aspiration was done and pus was sent for culture and sensitivity. The pus culture grew *Salmonella paratyphi A* which was sensitive to Ampicillin, Cefoxitin, ceftriazone, ceftazidime, ofloxacin, ciprofloxacin, amikacin, netrimycin, co-trimoxazole and gentamycin.

Antibiotics was changed and intravenous Ceftriaxone was started according to the sensitivity report. However, abscess collection recurred for which incision and drainage under local anaesthesia was done. She underwent regular saline dressings under antibiotic cover and the wound healed completely.

DISCUSSION

Salmonella paratyphi is one of the commonest infection in developing countries like India. Mostly it presents as enteric fever but rarely it can present as focal infection. The pathogenesis behind the localization is poorly understood. Some have hypothesized that post traumatic healed site can be a safe abode for the micro-organism to hide from immune system during the bacteraemia phase and slowly resulting in a localized infection.¹

There have been anecdotal reports of abscess in testis, kidneys, ovary, liver and breast.²⁻⁵ However, anterior abdominal wall abscess due to *Salmonella paratyphi A* is not yet reported. In rare cases dissemination to different sites of body where pre existing abnormality is seen in tissue or organ causing localized infection commonly seen in immunocompromised patients. This phenomenon has been reported in *Salmonella typhi* infection.^{1,6} Our patient had laparoscopic cholecystectomy 5 years ago. The healed fibrosed port site might have become a sanctuary for the

micro-organism during the bacteremia phase of febrile illness and gradually resulting in an abscess or it could have been a random seeding of the infection.

Antibiotic therapy alone is not enough and the abscess needs surgical or radiological drainage. Intravenous antibiotics are better than oral form and can result in faster recovery and prevent recurrence.⁷

CONCLUSION

Anterior wall abscess due to *Salmonella paratyphi A* is a very rare complication. Incision and drainage followed by appropriate antibiotics is the mode of management.

REFERENCES

1. Magdaline JM, Idikula MJ, Sebastian A. *Salmonella Typhi* causing parietal wall abscess. *J Acad Clin Microbiol.* 2014; 16: p. 30-1.
2. D'Cruz S, Kochhar S, Chauhan S, Gupta V. Isolation of *Salmonella paratyphi A* from renal abscess. *Indian J Pathol Microbiol.* 2009 Jan-Mar; 52(1): p. 117-9.
3. Navin PI, Thambu DS, Venugopal R, Subhash HS, Thomas K. *Salmonella paratyphi osteomyelitis and psoas abscess.* *Trop Doct.* 2006 Jan; 36(1): p. 58-9.
4. A.R. Jeansemail, M.W. McKendrickemail. *Salmonella Paratyphi A liver abscess—Secondary infection of an amoebic liver abscess?* *Travel Medicine and Infectious Disease.* 2007 March; 5(2): p. 144-146.
5. Nawaz GI, Rehman A, Muhammad S, Khawaja MA, Raja N, Aan N, Hussain I, Akhter S. *Testicular abscess caused by Salmonella para-typhi.* *J Ayub Med Coll Abbottabad.* 2011 Jul-sep; 3(3): p. 153-4.
6. Raghunath R, Ashok AC, Sridaran D, Indumathi VA, Behvadi MR. *Acute of injection abscess due to Salmonella Typhi.* *Indian J Med Microbiol.* 2003; 21: p. 59-60.
7. Shelanah Fernando, Janice Gail Molland, Thomas Gottlieb. *Failure of oral antibiotic therapy, including azithromycin, in the treatment of a recurrent breast abscess caused by Salmonella enterica serotype Paratyphi A.* *Pathogens and Global Health.* 2012; 106(6): p. 366-369.

Spinocerebellar Ataxia presenting with Signs of Amyotrophy:

A Case Report.

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Abstract : Spinocerebellar ataxia (SCA) is a progressive, degenerative, genetic disease involving cerebellum and its connections with multiple subtypes. It is an uncommon disease. We report a case of a 32-year male, who presented with bilateral cerebellar signs and amyotrophy with family history suggestive of an autosomal dominant pattern of inheritance. He was found to have spinocerebellar ataxia type 2 on genetic testing.

INTRODUCTION

Spinocerebellar ataxia (SCA) has a worldwide distribution, but some cases are more prevalent in one region than the other. SCA 2, SCA 3, and SCA 6 appear to be the most common and together account for nearly half of all families worldwide. SCA-2 is typically the most common among the SCAs in India,¹ and stands the next most common SCA after SCA-3

worldwide. Spinocerebellar ataxia type II (SCA 2) is characterized by gait and limb ataxia, dysarthria, ophthalmoplegia, and polyneuropathy.² Extrapyramidal signs and dementia are observed at late clinical stages. SCA 2 is caused by an expanded (CAG) trinucleotide repeat on the chromosome 12 resulting in production of abnormal protein called ataxin-2. The symptoms usually begin in the third or fourth decade of life.

In this paper we report about a family which was affected by SCA for three generations.

CASE REPORT

A 32 year old male, resident of Haryana, born out of nonconsanguineous marriage, with normal birth and developmental history, presented with imbalance while walking for 10

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years, with tendency to reel and fall in an unpredictable direction, along with in-coordination of both hands for same duration, causing difficulty in writing, eating, and operating appliances. The above symptoms were progressive in nature. There was a history of involuntary violent movements of lower limbs during sleep. His medical history was otherwise unremarkable. There was no relevant past history. There was no history of exposure to alcohol, drugs, or toxins. **There was history of similar symptoms in his grandmother and her brothers, his father and his uncle and daughter of his uncle, and his elder brother suggestive of autosomal dominant inheritance (Fig 1), however this could not be confirmed by molecular studies. The grandparents were affected in their sixties with mild ataxia without hampering their daily routine activities. Similarly, 2nd generation family members were also having ataxia with minimal functional impairment; however age of presentation was earlier than previous generation, whereas the age of onset in the proband and his siblings was even earlier with severe impairment.**

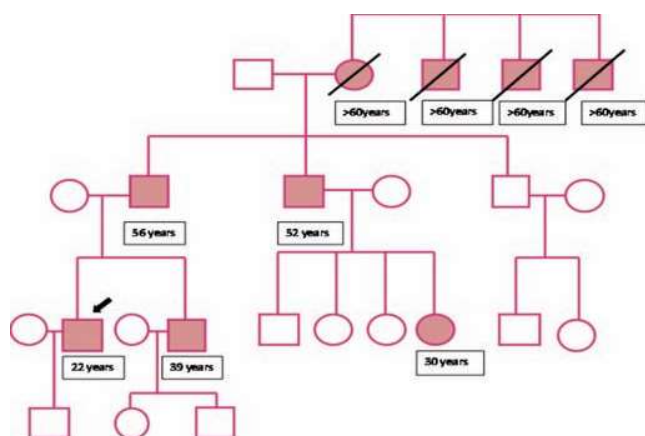


Figure 1: Pedigree showing Autosomal Dominant pattern of inheritance of SCA2 in three generations with approximate age of disease onset shown in boxes.

On Examination: He had normal vitals and general physical examination. His mental status was normal. Speech was dysarthric (scanning). He had lid retraction in right eye and slow horizontal saccades, however rest all cranial nerve examination was normal. No nystagmus was observed and both fundi were normal. Motor system examination revealed reduced bulk and atrophy of hands, power was 4/5 in all muscle groups, with decreased tone. Fasciculations, were present in tongue and all limbs. Superficial and deep tendon reflexes were absent in all four limbs, and plantars were flexor bilaterally. Joint position and vibration sense was reduced globally. There were cerebellar signs in both upper and lower limbs with ataxic gait.

Complete hemogram, liver function test, renal function test, thyroid function tests were normal. Serum Vitamin B12, folate levels were normal. No Kayser-Fleischer rings were seen on slit lamp examination. Chest X ray and Electrocardiogram were normal. Magnetic resonance imaging brain revealed olivopontocerebellar atrophy (Fig.2). Gene analysis revealed SCA2 positivity. Nerve conduction studies revealed decrease in conduction velocity and amplitude of bilateral Tibial, Peroneal and ulnar nerves suggestive of Neuropathy. Electromyography revealed that bilateral tibialis anterior, bilateral Vastus medialis muscles had 50-60% recruitment with mildly decreased amplitude; while bilateral deltoid showed 70-80% recruitment whereas bilateral abductor digiti minimi showed 20-30% recruitment with severely decreased amplitude. First dorsal interossei showed 5-10% recruitment. Motor unit action potentials (MUAPs) recorded were polyphasic with slightly decreased duration. Spontaneous fibrillation potentials as well as fasciculations were also recorded.



Figure 2: showing severe olivopontocerebellar atrophy.

DISCUSSION

Spinocerebellar ataxia type 2 represents a genetically defined neurodegenerative disorder characterised by autosomal dominant inheritance and progressive cerebellar ataxia, combined with slow saccades and sensorimotor neuropathy. The neuropathology comprises olivo-ponto-cerebellar atrophy (OPCA) with axonopathy of posterior columns, spinocerebellar tracts, and peripheral nerves. The underlying mutation of SCA2 consists of unstable expansion of the trinucleotide repeat (CAG)₈CAA(CAG)₄CAA(CAG)₈ within the ATXN2 gene exon 1 located on chromosome 12q24.1. This repeat encodes a polyglutamine (polyQ) tract in the protein ataxin-2. In normal individuals, the trinucleotide repeat length varies and contains between 13 and 27 units. Affected SCA2 individuals have 32 or more CAG repeats, with 37 to 39 repeats representing the most frequent pathologic expansion. The expanded alleles have lost interrupting CAA-triplets, a factor thought to promote the length instability. Currently, the function of ATXN2 is not clear, but several lines of evidence evoke its involvement in RNA metabolism. ATXN2 and its orthologues in other organisms relocalize during periods of cellular stress to mRNP granules where mRNA is stored during translation repression, promote the formation of these stress granules and inhibit cell growth.

Mean age at onset is typically in the fourth decade, however Anticipation may occur, particularly with paternal transmission; those affected individuals generally have longer CAG repeat lengths and earlier symptom onset age. SCA2 has been reported in various ethnicities including Cuban, Indian, Italian, Mexican, South African, and Spanish. Besides ataxia, SCA2 features may include slowed saccades (which may progress to ophthalmoparesis), brisk deep tendon reflexes (which may progress to areflexia), peripheral neuropathy, dementia, myoclonus, dystonia, chorea, and levodopa-responsive parkinsonism. Sleep disturbances are frequent complaints of SCA2 patients and their relatives. The most prominent sleep disorders are restless legs syndrome. Milder phenotypes with less prominent ataxia, neuropathy, dystonia, and myoclonus but greater parkinsonian features have been associated with shorter CAG repeat expansions.

In the reported family, affected members had cerebellar dysfunction of variable degree. Most patients began with dysarthria and gait ataxia. As the disease progressed, limb ataxia became more pronounced. Family members who were examined showed slowness of horizontal saccadic eye movements to a variable degree and limb and gait ataxia, but none of the family members were affected so early and with severe degree of impairment (inability to carry out daily routine activities) as the index case. These findings could be attributed to the phenomenon of anticipation and different degrees of expansion in maternal or paternal transmission. The clinical suspicion of SCA type 2 was confirmed by genetic study in two members (patient and his uncle). Secondly, none of the family members have the fasciculations and features of amyotrophy except for the index case. These findings were suggestive of possibility of motor neuron disease (MND) in the present case. Although the patient did not exhibit any upper motor neuron signs on presentation, the presence of generalized muscle weakness, atrophy and fasciculations classically involving the tongue and generalized body (more in