Letter to Editor

Enthesitis-Related Arthritis in a Child with Turner Syndrome

Dear Editor,

A 12.5-year-old girl had a history of subacute back and left hip pain since 11 years of age. Old records revealed clinically demonstrable sacroiliitis and left hip arthritis. She was HLAB27 positive and was diagnosed as enthesitis-related arthritis (ERA). She had received 8 weeks of medium-dose oral steroids and was subsequently well-controlled on oral sulfasalazine. Her course had been punctuated by two episodes of acute anterior uveitis treated with topical steroids. Eighteen-months later, the family sought multidisciplinary-care for her rheumatology follow-up and short stature at our center.

Review of previous growth-records revealed short stature even at age 10 years, prior to the onset of arthropathy. Her parents mentioned her being "much shorter than her peers" throughout schooling. The family history was not contributory. Certain syndromic clues were noted; palatal nevus, absence of thelarche denoting delayed puberty and bilateral cubitus valgus [Figure 1a and b]. The initial evaluation with a pelvic sonogram revealed hypoplastic uterus and streak ovaries. G-banded karyotype depicted monosomy of X-chromosome (45, ×0) confirming turner syndrome (TS). Screening for associated comorbidities (blood-pressure, 2D-Echo for aortic-aneurysms and aortic-dissection, electrocardiogram, renal-sonogram, audiogram, comprehensive ophthalmic examination, laboratory tests to rule out dysglycemia, thyroiditis, celiac disease, and dyslipidemia) returned normal. After appropriate endocrine work-up, she received daily recombinant GH-injections (40 mcg/kg/day) and documented a height spurt of 14-cm within the following 18-month. Puberty induction was delayed intentionally after due discussion to optimize GH-related height gain. Her ERA continued to be under control on sulfasalazine. Follow-up surveillance based on standard TS guidelines was advised.

The association of TS with spondyloarthropathy (ERA in our case) has been reported only four times before.[1-4] Haploinsufficiency of certain "protective" X-linked genes is postulated for the occurrence of male-preponderant ERA in girls with TS.[4] Our case is unique in also highlighting the impact of timely diagnosis through timely GH treatment and targeted multidisciplinary surveillance.

Rheumatologists caring for children and adolescents, may encounter short stature attributable to multiple mechanisms in their practice, namely, underlying chronic inflammatory process (e.g., juvenile idiopathic arthritis), long-term steroids, innate to the disease process (e.g., autoimmune thyroiditis, celiac disease), or an associated comorbidity or syndrome as in our case. Notably, TS is

Figure 1: (a) Frontal picture of the child. Note bilateral cubitus valgus. (b) Open mouth depicting palatal nevus

the most common cause of short stature in a female after excluding familial and constitutional short stature.[1]

TS is associated with a 2–3-fold increase in autoimmune and rheumatological diseases. These individuals could seek rheumatology services for multiple reasons; common among them being autoimmune thyroiditis, celiac disease, type-I-diabetes mellitus, rheumatoid arthritis, inflammatory bowel disease, and psoriatic arthropathy.[5] All these diseases may out-shadow the subtle forms of TS and antedate its diagnosis. The absence of physical clues of TS or attributing the short stature to coexisting factors, mimics or steroid therapy, are common pitfalls. Detailed inspection of retrospective growth charts and thorough clinical examination are crucial, as exemplified by our case.

"Heightened" awareness about TS in rheumatologists can positively impact timely GH therapy and puberty induction and consequent bone mass, final adult height, and reproductive function; besides planning surveillance for cardiovascular complications, and other comorbidities associated with TS.[1,6]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the parents have given their consent for images and other clinical information to be reported in the journal. The parents understand that names and initials will not be published, and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

AQ1 Growth hormone

AQ2: Yes, should be number 5

AQ3: Approve. We have consent form available

AQ7: Ref number 6, should be renumbered as 5

Conflicts of interest

There are no conflicts of interest.

Shruti Bajaj, Prashant Patil, Raju Khubchandani

AQ4 *???, NH SRCC Children's Hospital, Mumbai, Maharashtra, India*

AQ4: Section of Clinical Genetics, Pediatric Endocrinology and Pediatric Rheumatology

Address for correspondence: *Dr. Shruti Bajaj, 403, Mickey's Paradize, Opposite Orlem Garden, Tank Road, Orlem, Malad (West), Mumbai - 400 064, Maharashtra, India. E-mail: drshru.a@gmail.com*

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AQ7

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Author Queries???

AQ1: Kindly provide expansion.

AQ2: As required by the journal style, reference citations in the text have been arranged in order to appear in sequential order. Please check.

AQ3: Based on the information available in FP document that consent form has been obtained from all patients we have included the declaration statement.

AQ4: Kindly provide department.

AQ5: Please provide revised date

AQ6: Please provide accepted date

AQ7: Kindly provide 5th reference details