then spread to the entire body including limbs. It was itchy. The family are originally from Quebec and had been residing in Ireland for the past 2 years in relation to the father’s occupation. Contact with infectious disease and recent travel were denied. He had never been hospitalised and had no medical diagnoses. His mother was adamant that all vaccinations were up to date and included the Varicella vaccine at one year of age in Canada. He had no known allergies and was not taking any medications.

On examination he had multiple vesicular and pustular lesions with an erythematous base. Some lesion had necrotic centres. There were some vesicles on his lower lip, buccal mucosa and also on the throat.

His WCC was 2.08, Neutrophils 0.73, Lymphocytes 0.73, CRP 20. Influenza, RSV and Monospot were negative.

He was initially treated with IV Augmentin and Fluoxacillin for a presumed diagnosis of Impetigo. More lesions appeared over his trunk and abdomen over the subsequent 24 hours although he was not systemically unwell. The Dermatologist made a clinical diagnosis of ‘Chicken Pox’. He was discharged home on an immunocompetent dose of oral acyclovir pending results of skin swab, throat swabs and Varicella titre.

At follow up one week later he was clinically well with multiple healing lesions. His Varicella titre were high confirming a diagnosis of Varicella Zoster infection. His mother brought with her his vaccination records from Canada which showed that he had received only one dose of the Varicella vaccine and had missed the booster. This is in contrast to his siblings who were fully vaccinated and did not develop Varicella despite close contact.

Conclusion Common conditions are common. Vaccines can fail. Parents should be encouraged to keep detailed records of all vaccinations including boosters and to follow through with booster vaccines when indicated.

P225 CONGENITAL HYALINE FIBROMATOsis SYndrome: A CASE REPORT OF HETEROZYGOUS ANTXR2 AND LITERATURE REVIEW

Monica Salama*, Sarah Kash, Isabelia Gover, Abdalla Hamza. Mayo University Hospital, Castlebar, Ireland

Introduction Juvenile hyaline fibromatosis is a rare hereditary disease characterized by deposits of a clear substance (hyaline) in the skin and other body tissues, it becomes apparent at birth or in infancy presenting with severe pain with movement, skin lesions & bumps , gingival hyperplasia, progressive joint contractures, and bone lesions.1 Complications can be life threatening. The inheritance follows autosomal recessive pattern, Causing mutations in the ANTXR2 gene. Treatment is supportive and aims to alleviate pain and other symptoms of the condition. The prevalence is unknown; about 84 people with this disorder have been reported in 2018 ².

Case We report a 23 month old boy, born at 38 weeks gestation, no neonatal complications. He was referred to paediatric clinic at 3 month old with excessive crying, feeding difficulty and failure to thrive crossing down on centiles from 9th to <0.4th centile. At 8 months noted to have limited movement of both shoulders. He then started to have gingivitis and gum swelling which progressed to gingival hyperplasia. Developmentally he sat unsupported at 8 months, not crawling or standing no concerns about hearing or vision , plays and interacts well with siblings.

Parents are non consanguineous of Syrian origin, He has two sisters and one brother all well. They had no skin lesions or joint problems. There was no history of similarly affected relatives.

Physical examination He appears symmetrically small, generally bright visually alert to his surroundings. Visible nodules over left ear and left nostril, gum hypertrophy .Decreased muscle bulk in his deltoids with limited passive movement of both shoulders and extreme pain on any shoulder movement. Cannot stand or walk independently.

Laboratory investigations Biochemical and metabolic work up was all normal DNA PCR: Heterozygous ANTXR2 gene detected.