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Commentary

Significance of Coronary Artery Micro-Fistulas in Kleefstra Syndrome

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DESCRIPTION

Kleefstra syndrome (OMIM 610253) (KS) is a rare genetic condition also known as 9q subtelomeric deletion syndrome. Although the prevalence rate is unknown, a number of additional instances have been diagnosed since the widespread use of subtelomeric FISH and, later, array Comparative Genomic Hybridization (aCGH). In the syndrome developmental delay/intellectual impairment phenotype, all individuals have brachy(micro)cephaly, unusually formed brows, flat face with hypertelorism, short nose with anteverted nostrils, thicker lower lip, carp mouth with macroglossia and infantile hypotonia. Additional symptoms such as cardiac abnormalities, microcephaly, genital/renal abnormality, recurrent infections, hearing impairment, and tracheo/bronchomalacia are seen in a subset of individuals. Epilepsy and psychological issues are both significant and prevalent symptoms.

While typical drugs may generally treat epilepsy, mental abnormalities might include apathy, violent episodes, psychosis, autistic traits, bipolar mood disorders, and regression in the daily function and cognitive ability. Furthermore, less common symptoms noted include micropenis, cryptorchidism, and vesicouretheral reflux. The majority of patients have a submicroscopic deletion in the chromosomal region 9q34.3 or disruption of the *Euchromatin Histone Methyl Transferase 1 (Eu-HMTase1)* gene, which results in haploinsufficiency of the *EHMT1* gene. So far, no genotype-phenotype association with deletion size or between patients with deletions and those with mutations has been discovered. Coronary Artery Fistulas (CAF) are uncommon coronary circulation system abnormalities. They are congenital or acquired abnormalities of the coronary artery assembly and circuitry. Coronary artery fistulas are classified into two types. Coronary-cameral fistulas are abnormal connections that occur between the coronary arteries and any of the heart chambers. Coronary arterio-venous malformations are anomalous connections between coronary arteries and systemic/pulmonary circulation vessels. Although the majority of coronary artery fistulas are discovered by chance during a coronary catheterization, a subset of patients with this disease may exhibit signs and symptoms of CHF (Congestive Heart Failure), MI (Myocardial Infarction) and other cardiopulmonary functional abnormalities. In terms of diagnosis, coronary angiography and coronary Computed Tomography Angiography (CTA) are regarded as extremely accurate methods for detecting coronary artery fistulas. Catheterized closure is often recommended as the primary treatment approach.

Children may experience hemodynamically significant issues as a result of coronary artery fistula, which is characterised as an improper interaction between the coronary arteries and a cardiac chamber (most frequently) or a thoracic great vessel. Children with coronary artery fistulas may be evaluated using cardiac catheterization, cardiac MRI, and cardiac CT. For the precise diagnosis of coronary artery fistula in children, CT has been used crucially. Hemodynamically significant coronary artery fistulas require surgical or interventional therapy.

CONCLUSION

Depending on the size of the channel, CAF is a rare aberration with a varied clinical course that can range from no symptoms to major problems like heart failure and myocardial infarction.

Accurate evaluation of the complex physiology of CAFs, including the location and number of sources and drainage sites, as well as any related defects, can be accomplished using ECG-gated CT angiography with 3D reconstruction. Radiologists need to be completely aware of the crucial role that CT angiography plays in the assessment of CAFs in order to help clinicians in making the best clinical and therapeutic decisions.

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