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Congenital Midline Cervical Cleft

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ABSTRACT

Congenital midline cervical cleft is a rare anomaly that typically presents in the neonatal period as a thin suprasternal vertical band of erythematous skin with a nipple-like projection superiorly, which may exude fluid. We present the clinical and pathophysiologic features and the imaging findings of this uncommon, and rarely described entity in a newborn girl.

CASE REPORT

CASE REPORT

A large for gestational age female was born to a healthy gravida 6 para 5 woman at 41 weeks of gestation. The prenatal course was unremarkable and, although at birth the infant was found to have meconium stained amniotic fluid, there were no neonatal complications. She was transferred to our institution for further evaluation of a one centimeter vertically-oriented linear band of erythematous, atrophied skin in the suprasternal neck with a 2 mm nipple-like protrusion cranially and a blind-ending sinus tract caudally (Figure 1). No other dysmorphic features were present and there was no family history of congenital abnormality or consanguinity.

Initial radiologic evaluation with a targeted ultrasound to the soft tissues of the neck demonstrated an apparently blind-ending midline suprasternal sinus tract arising from the subcutaneous soft tissues (Figure 2).

Given the infant's stridor on exam however, there was persistent concern for a fistulous connection of the sinus tract. Rigid bronchoscopy revealed an intact trachea without fistulous communication or evidence of tracheomalacia.

A targeted magnetic resonance imaging examination was performed at 3 tesla (Discovery, GE Healthcare, Waukesha, WI) including T2 with and without fat saturation and pre- and postgadolinium enhanced T1-weighted sequences. This demonstrated a T1 hypointense, T2 hyperintense tract with peripheral post-contrast enhancement at the base of the neck measuring 7 mm in craniocaudal length and 3 mm in width. This tract extended from the skin surface at the inferior aspect of the lesion to the superior cortex of the sternal manubrium in a caudal direction. There was no cortical disruption or involvement of the manubrium. Additionally, although the tract was noted to course toward the thyroid gland, the thyroid gland appeared separate and intact. There was no associated soft tissue mass or fluid collection (Figure 3). These MRI findings confirmed the diagnosis of a congenital midline cervical cleft.

The neonate continued to do well and, after thorough evaluation by pediatric surgery, was discharged home with scheduled outpatient follow-up for surgical management of the congenital midline cervical cleft.

DISCUSSION

Etiology and demographics

Congenital midline cervical cleft is a rare entity that was first described in the English surgical literature by Bailey in 1924. Subsequently there have been fewer than 50 case reports in the English pediatric surgical literature documenting this congenital anomaly [1]. To date this entity has not been reported in the radiologic literature. The exact incidence is unknown, however, it comprises up to 2% of all neck anomalies and appears to be more common in Caucasian females [2,3].

Although the embryology of the head and neck is thought to be well understood, the pathogenesis of congenital midline cervical clefts remains speculative. The most widely accepted explanation for the development of this anomaly is impaired fusion of the first or, more commonly, second branchial arches in the midline as well as improper interaction between the ectoderm and mesoderm [4]. In normal embryogenesis the branchial arches grow medially and merge cephalad to caudal. Prior to fusion, mesodermal tissue migrates between the arches and pushes ectoderm outward to flatten the ventral furrow [1,2]. In the setting of congenital midline cervical clefts some mechanical factor or vascular anomaly is thought to result in ischemia or necrosis and interfere with these normal processes. Other less accepted explanations include increased pressure in the cervical region from the developing pericardial roof, amniotic adhesions, or exteriorization of a thyroglossal duct cyst. There are no associated risk factors [5].

Histologic examination has revealed stratified keratinized squamous epithelium lacking skin appendages such as sweat or sebaceous glands. The dermis has alternating hypertrophy and atrophy and there is a mild lymphocytic inflammatory infiltrate in the subcutaneous tissues [1]. An associated sinus tract is lined by a pseudostratified epithelium with seromucinous salivary glands; however, there have been several reports of respiratory epithelium and bronchial glands associated with the sinus tract [2].

Clinical and imaging findings

The midline cervical cleft is usually detected at birth as a thin vertical band of erythematous skin with a nipple-like projection superiorly. This lesion can be found anywhere along the midline from the chin to the suprasternal notch. An associated blind-ending sinus tract in the caudal aspect may occasionally have minimal secretion. The midline cervical cleft heals by scarring and resultant fibrous cord formation that may lead to a neck contracture and eventually torticollis or micrognathia in early childhood [3]. Additionally, there have been reports of associated lower lip, mandible, chin, and tongue defects [1].

Prior to the widespread use of ultrasound and MRI, radiographs of the neck classically demonstrated bony spurring at the mandible in advanced cases due to traction from the underlying fibrous cord [4]. Ultrasound of the neck shows a non-vascular, blind-ending sinus tract from the skin surface. The thyroid gland is normal. MRI is helpful in defining the lesion and its course as well as to exclude other differential considerations such as thyroglossal duct cysts or branchial cleft anomalies. MRI usually demonstrates skin thinning and a peripherally-enhancing, T1 hypointense, and T2 hyperintense linear tract without disruption of any bony or cartilaginous structures or involvement of the thyroid gland.

Treatment and prognosis

Treatment is accomplished surgically, by resection of the skin tag and fibrotic tissue with closure of the soft tissue defect by Z-plasties. Although there is no consensus on age of treatment, surgical intervention is recommended early before scarring leads to contracture, neck webbing, or mandibular hypoplasia [1]. Multiple series have documented good outcomes, however, there have been reports of recurrence [3].

Differential diagnosis

Differential diagnosis includes thyroglossal duct cysts or branchial cleft anomalies. A thyroglossal duct cyst that, although a midline T2 hyperintense lesion that may have faint peripheral enhancement, characteristically appears cystic and it is typically much higher in the neck, being more midline when related to the hyoid and more lateral when in the infrahyoid neck. In addition, a thyroglossal duct cyst does not usually have an associated sinus tract unless it is superinfected [1,2,5]. 2nd or 3rd branchial cleft cysts or pyriform sinus fistulae are typically off midline in the lateral neck [6].

TEACHING POINT

The congenital midline cervical cleft is a rare entity that is usually diagnosed clinically, however, the radiologist may be asked to evaluate this anomaly to further characterize anatomy and exclude other differential considerations. Recognition of an anterior cervical midline skin defect with an underlying blind-ending sinus tract that is T1 hypointense, T2 hyperintense, and demonstrates peripheral contrast enhancement on MR allows early surgical intervention to prevent future complications.

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FIGURES

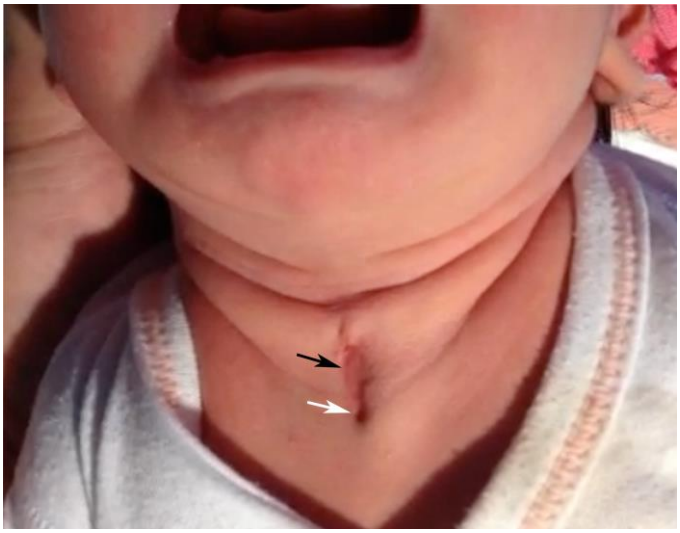
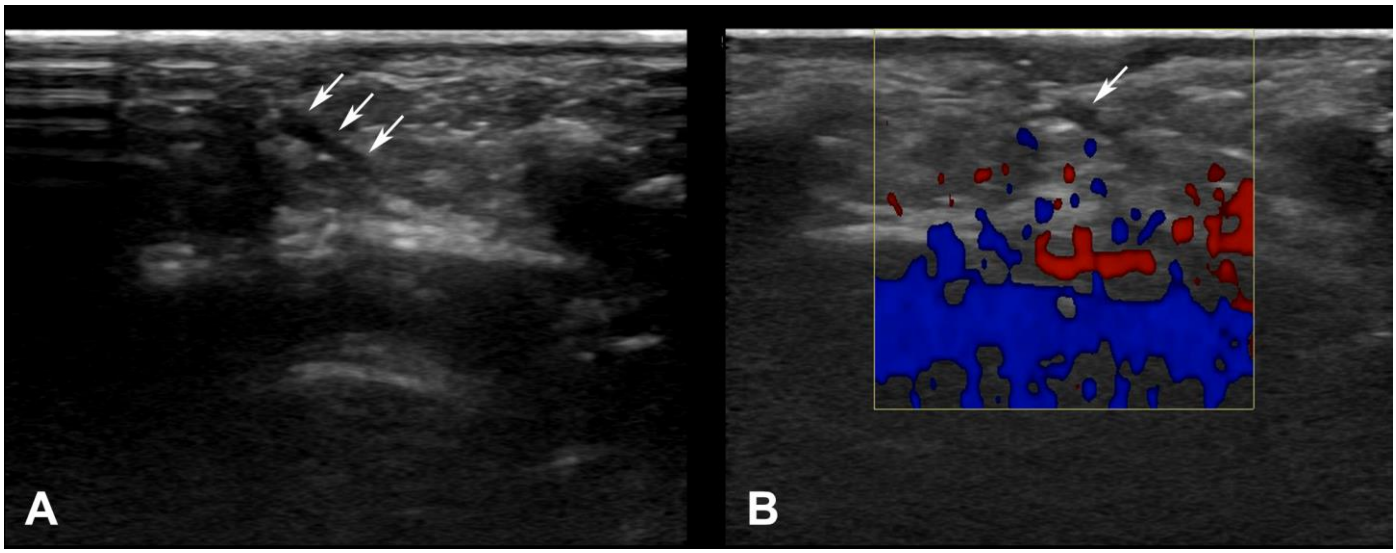


Figure 1 (left): A two-day old girl with congenital midline cervical cleft. FINDINGS: Clinical photograph of a 2-day old girl shows a band of thin, erythematous skin (black arrow) in the midline of the lower neck with a sinus tract caudally (white arrow).

Figure 2 (bottom): A two-day old girl with congenital midline cervical cleft. FINDINGS: Transverse US (A) and longitudinal color Doppler US (B) images demonstrate a blind-ending, caudally-oriented tract (white arrows, A) without associated vascularity. TECHNIQUE: Transverse and longitudinal scan planes of a neck US (A and B). Ultrasonography was performed using an 18 MHz linear transducer (GE Logiq).



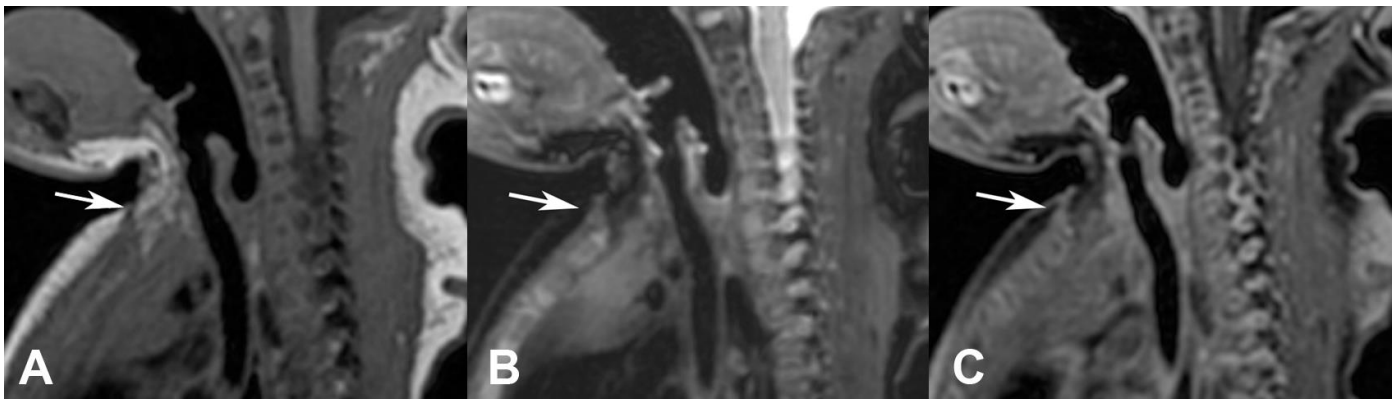


Figure 3: A two-day old girl with congenital midline cervical cleft. FINDINGS: Magnified sagittal T1-weighted (A), fat-saturated T2-weighted (B), and post-gadolinium fat-saturated T1-weighted (C) images show a T1 hypointense, T2 hyperintense, and peripherally-enhancing cranio-caudally oriented tract (white arrow). The tract courses from the skin in the midline at the base of the neck toward the manubrium of the sternum. TECHNIQUE: 3.0 Tesla MRI (GE Discovery) Image A: Sagittal T1 TR/TE - 600/8 ms, 1.5 mm slice thickness, noncontrast; Image B: Sagittal T2 TR/TE - 3000/62 ms, 1.5 mm slice thickness, noncontrast; Image C: Sagittal T1 TR/TE - 600/8 ms, 1.5 mm slice thickness, Gadolinium contrast 0.2 mL/kg.

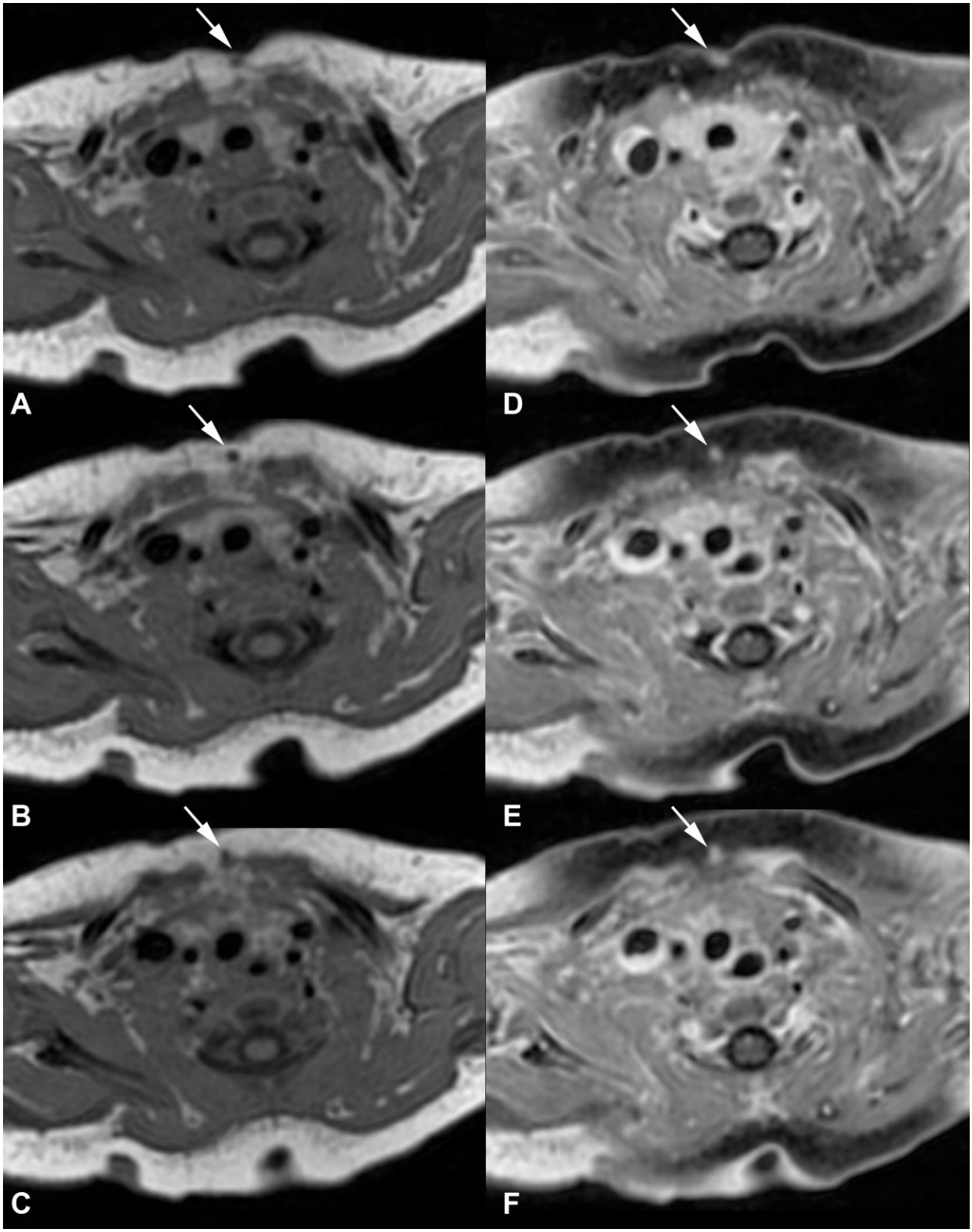


Figure 4: A two-day old girl with congenital midline cervical cleft. FINDINGS: Serial axial T1-weighted images (A-C) show a hypointense tract (white arrows) that enhances on corresponding axial post-gadolinium T1-weighted images with fat saturation (D-F). The tract extends cranio-caudally from the skin surface toward the manubrium of the sternum. TECHNIQUE: 3.0 Tesla MRI (GE Discovery) Images A, B, C: Axial T1 TR/TE - 600/8 ms, 1.5 mm slice thickness, noncontrast. Images D, E, F: Axial T1, TR/TE - 600/8 ms, 1.5 mm slice thickness, Gadolinium contrast 0.2 mL/kg.

Etiology	Uncertain, several hypotheses proposed, the most accepted is impaired branchial arch fusion. Others include increased pressure from the developing pericardial roof, amniotic adhesions, or exteriorization of a thyroglossal duct cyst
Incidence	Extremely rare, seen in up to 2% of all congenital neck anomalies
Gender Ratio	Female predominance
Age Predilection	Congenital, detected at birth
Risk Factors	Not known
Treatment	Surgical resection
Prognosis	Favorable, with some reports of recurrence
Imaging Findings	Radiographs may show bony mandibular spur from traction. Blind ending sinus tract on ultrasound. MRI demonstrates skin thinning and a peripherally-enhancing, T1 hypointense, and T2 hyperintense linear tract

Table 1: Summary table for congenital midline cervical cleft.

	X-Ray	**US**	**CT**	**MR**
Congenital Midline Cervical Cleft	Occasionally see bony mandibular spur	Midline blind-ending sinus tract	Not routinely imaged, blind ending sinus tract	T1 hypointense,T2 hyperintense tract with peripheral enhancement
Thyroglossal duct cyst	Usually no findings	Midline anechoic or hypoechoic cyst near hyoid bone.	Low-attenuating cyst. Mild peripheral enhancement	T1 hypointense, T2 hyperintense cyst. Wall enhances if superinfected
2nd Branchial cleft anomaly	Usually no findings	Anechoic or hypoechoic cyst posterolateral to submandibular gland and anterior to sternocleidomastoid	Low-attenuating cyst, nonenhancing wall, may enhance if superinfected	T1 hypointense, T2 hyperintense cyst. Wall enhances if superinfected

Table 2: Differential diagnosis table for congenital midline cervical cleft.

ABBREVIATIONS

CT = computed tomography
MRI = magnetic resonance imaging
TE = echo time
TR = repetition time
US = ultrasound

KEYWORDS

Congenital midline cervical cleft; midline cervical cleft; congenital midline cervical anomalies; branchial arch defects; pediatrics

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