New Association between Idiopathic Scoliosis and Luckenschadel Skull (Lacunar Skull)

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Highlights of the Study

- Luckenschadel skull is considered a rare radiological finding associated with severe neurological abnormalities.
- We found that the lacunar skull is a very common benign condition that is not associated with any additional neurological pathology in children with idiopathic scoliosis. In addition, it does not disappear over time; in fact, it may become more obvious even up to the age of 16 years.

Keywords
Luckenschadel skull (lacunar skull) · Idiopathic scoliosis · Chiari malformation

Abstract
Objective: Luckenschadel skull is a skull that is radiologically characterized by lacunae in the cranial vault. To date, although the association between neurological abnormalities and scoliosis is well recognized, no relationship between idiopathic scoliosis and a lacunar skull has been defined. We explored the incidence and time courses of lacunar skulls in patients with idiopathic scoliosis. Materials and Methods: Spinal X-rays of 3,170 children aged 6–16 years with idiopathic scoliosis evaluated from October 2010 to August 2020 were examined for the presence of an irregular inner calvarial table indicative of a Luckenschadel skull. A total of 1,760 (55.5%) of the 3,170 images included the skull. We also explored the frequency of intraspinal abnormalities in children with lacunar skull images who underwent spinal magnetic resonance imaging. Results: The study population consisted of 1,760 children, 1,203 girls (68.4%) and 557 (31.6%) boys. A total of 268 (15.2%) clearly evidenced lacunar skulls in their radiographs; 186 (69.4%) girls (11.3 ± 4.3 years) and 82 (30.6%) boys (12.6 ± 3.3 years). Two of the 56 patients (3.6%) who underwent spinal MRI had intraspinal abnormalities (isolated Chiari malformation-I). No additional neurological problems were detected in children with lacunar skulls. Conclusion: We conclude that the lacunar skull is very common in children with idiopathic scoliosis who lack any other neurological pathology. The lacunar skull does not disappear even in adolescence. Although previous publications have stated that lacunar skull disappears over time in radiographic images, we observed that it became more noticeable over time in children with scoliosis.

Introduction

Luckenschadel skull, also known as the lacunar skull or "cranialacunae," is a dysplasia of the membranous skull vault and is readily apparent in plain radiography. It is a congenital defect, the physiopathology of which re-
malformations [12]. Dauser et al. [13] were the first to describe the diagnosis and follow-up of patients with scoliosis has been extensively studied with more severe curves (>40°) [11].

The prevalence and severity of scoliosis are higher in girls than boys, with the female:male ratio increasing from 1.4:1 in prepubescent children to 7.2:1 in patients with mild curves (10–20°) to 7.2:1 in patients with severe curves (≥20°) [11].

In recent years, the routine use of full spinal MRI for diagnosis and follow-up of patients with scoliosis has greatly increased the detection rate of neuro-axis abnormalities [12]. Dauser et al. [13] were the first to describe the association between the CM and scoliosis, and the literature has grown over the past 3 decades. Intraspinal neural axis abnormalities such as an isolated CM, isolated syringomyelia, a CM combined with syringomyelia, and a tethered cord combined with diastematomyelia are common among patients with idiopathic scoliosis [14–17]. The prevalence of such conditions has ranged from 11.1% to 26.0% in studies with small sample sizes [18]. A recent review showed that the prevalence of neural axis anomalies in children with idiopathic scoliosis is estimated to be around 11% [19]. Scoliosis commonly coexists with the neural axis anomalies, but no causal relationship has yet emerged.

To the best of our knowledge, no study has yet sought an association between a lacunar skull and scoliosis. All prior reports have linked lacunar skull to only severe neurological abnormalities. Thus, we explored the frequency of lacunar skull in children with idiopathic scoliosis by evaluating a large number of radiographic images.

Materials and Methods

The Bezmialem Vakif University Local Research Ethics Committee approved this retrospective study and waived the requirement for written informed consent. We interrogated the database of Bezmialem Vakif University Hospital (Istanbul) to identify lacunar skull in children diagnosed with idiopathic scoliosis from October 2010 to August 2020. The medical records of 3,170 children aged 6–16 years with lateral spinal curvatures (Cobb angles) ≥10° who underwent spinal X-ray were reviewed by the same observer in terms of the presence of an irregular inner calvarial table indicative of Luckenschadel skull. The study population consisted of 1,760 children with scoliosis: 1,203 girls (68.3%) and 557 (31.6%) boys. A total of 284 (15.2%) clearly exhibited lacunar skulls in spinal radiographs. Of these, we studied 268 (15.2%) children: 186 (69.4%) girls and 82 (30.6%) boys, with a female: male ratio of 2.26:1. The mean ages were 11.3 ± 4.3 years for girls and 12.6 ± 3.3 years for boys. In the present study, Luckenschadel skull was defined by groups of round or oval clearly evidenced finger-shaped pits on the inner surface of the vault, which vary in size, shape, and number, separated by ridges of the bone. We also evaluated the presence of intraspinal abnormalities in children with scoliosis for whom lacunar skull images were available and who underwent spinal MRI, by examining radiology reports.

Subjects

We enrolled a large group of subjects to determine the frequency of lacunar skull in children with scoliosis diagnosed via standing spinal radiography. Children with infantile scoliosis (age at onset <3 years) and with scoliosis that commenced at <6 years often exhibiting congenital neurological anomalies were excluded. We screened the records of idiopathic scoliosis patients aged 6–16 years. Scoliosis was defined as a lateral spinal curvature (a Cobb angle) ≥10°. A total of 3,170 patients were initially included. A total of 1,760 (55.5%) of the 3,170 images included the skull. We excluded 16 children with congenital neurological diseases or congenital scoliosis.
**Inclusion/Exclusion Criteria**

The inclusion criteria were children aged 6–16 years at diagnosis; scoliosis (Cobb angle ≥10° in coronal images) as noted by radiologists; children with scoliosis whose skulls were imaged via anteroposterior, standing full-spine radiographs; and with initial normal neurological findings on medical records. The exclusion criteria were congenital scoliosis and any neuromuscular condition or syndrome. Chiari malformation type I (CM-I) was defined radiographically as a simple displacement of the cerebellar tonsils 5 mm or greater below the foramen magnum on MRI.

**Data Analyses**

All analyses were performed using the IBM Statistical Package for the Social Sciences for Windows (ver. 20.0; IBM Corp., Armonk, NY, USA). The results are presented as means ± standard deviations for continuous variables and as frequencies with percentages for categorical variables. A \( p \) value <0.05 was taken to indicate statistical significance.

**Results**

The study population consisted of 1,760 children, 1,203 girls (68.4%) and 557 (31.6%) boys. A total of 268 (15.2%) clearly evidenced lacunar skulls in their spinal radiographs; 186 (69.4%) girls (11.3 ± 4.3 years) and 82 (30.6%) boys (12.6 ± 3.3 years). The lacunar image was mostly detected in the frontal and parietal bones. The female sex was significantly associated with scoliosis (68.3%) and lacunar skull (69.4%) in spinal radiographs (both \( p < 0.05 \)). Of the 268 children enrolled in the study, 151 (56.3%) were adolescents aged 12 years and over. The study group consisted of 117 (43.7%) children under 12 years of age. We noted that 142 (52.9%) of 268 patients had been followed in our hospital for at least 3 years, and the lacunar head image persisted until the last radiographs. We observed that the lacunar head image of 29 (10.9%) children became more pronounced with age, regardless of the Cobb angle. Although previous reports have stated that the lacunar skull became less apparent over time, we observed that the skull persisted over time, often became more obvious, and was still present at the age of 16 years (shown in Fig. 1–4).

At our institution, full spinal MRI was performed more often in patients with early-onset scoliosis (before corrective surgery) to evaluate the possible presence of an underlying osseous or neurological problem. Full spinal MRI was not routinely performed on all patients with idiopathic scoliosis. Therefore, most of the 268 patients did not undergo MRI; 56 underwent full spinal MRI. Two patients (3.6%) exhibited intraspinal neural axis abnormalities in MRI. One had isolated CMs-I (9-year-old girl), and one had isolated CMs-I (11-year-old girl). The ages at first scoliosis diagnosis were 9 and 8 years, respectively (shown in Fig. 5, 6). No additional neurological problems were found in the medical records of the remaining 266 patients with lacunar skulls. During the 10-year period, 19 of the 268 patients with such skulls underwent operative correction of their scoliosis.

**Discussion**

The associations between intraspinal neural axis abnormalities and idiopathic scoliosis are well known, but no relationship has been identified between idiopathic scoliosis and the lacunar skull. We found, contrary to
common belief, that the lacunar skull is a very common benign condition that is not associated with any additional neurological pathology in children with idiopathic scoliosis. Two hundred sixty-eight of 1,760 patients with idiopathic scoliosis exhibited clear lacunar skulls in spinal radiographs; 56 of them underwent full spinal MRI and 2 had isolated Chiari malformation-I. Neither of the 2 patients with a CM-I required surgery.

The term “lacunar skull” refers to a dysplasia of the membranous bone; well-defined radiolucent areas in the calvaria correspond to nonossified fibrous bone. Lacunar skull has been frequently (up to 80% of cases) associated with CMs I–II and less commonly with encephalocele and craniosynostosis [20–22]. Coley et al. [23] performed neurosonography on 21 patients with myelomeningoceles to explore the possible presence of an irregular inner

Fig. 2. A 15-year-old adolescent boy with marked lacunar skull (multiple oval, round, and finger-shaped pits on the inner surface of the vault, lie in frontal and parietal bones) and idiopathic scoliosis (right thoraco-lumbar scoliosis, curve of 29°) without any additional neurological pathology.

Fig. 3. A 13-year-old adolescent girl with marked lacunar skull and idiopathic scoliosis (right thoracic left lumbar scoliosis: respectively, curve of 45° and 40°).
calvarial table indicating Luckenschadel skull; 9 patients exhibited prominent Luckenschadel findings, 9 had mild findings, and 3 were normal. The condition was first described by Engstler [3] as the “Luckenschadel” (gap skull) of a 4-day-old infant with spina bifida and has since been considered to reflect a severe neurological disease. To the best of our knowledge, this is the first report to show that lacunar skull is very common in children with idiopathic scoliosis lacking any neurological problem.

This condition has been suggested to be a fetal developmental anomaly caused both by changes in intracranial pressure during pregnancy and developmental anomalies of the calvarium and internal periosteum [24]. A current theory suggests that the CM-II and the Luckenschadel skull reflect the absence of cerebral ventricular distension because of decompression of an open neural tube [25]. Such a lack of stretching inhibits the normal development of membranous plates of the fetal calvarium, creating abnormally irregular collections of collagen fibers that, when ossified, yield the radiographic findings of the Luckenschadel skull. Its dissolution at 6 months of age may reflect remodeling in response to the normal enlargement of cerebral tissue or the development of hydrocephalus [26]. To the best of our knowledge, this theory has not yet been proven, and we believe that it is incorrect. In our study, the lacunar skull was found to persist even

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Fig. 4. A 16-year-old adolescent girl with marked lacunar skull and idiopathic scoliosis (right thoracic scoliosis, curve of 20°).

Fig. 5. A 9-year-old girl diagnosed idiopathic scoliosis (left lumbar scoliosis, curve of 22°) and lacunar skull. MRI scan demonstrated a CM-I (tonsillar herniation into the foramen magnum, 6.81 mm).
in adolescents with idiopathic scoliosis but without any underlying neurological disease. We believe that Luckenschadel skull is not attributable to increased intracranial pressure, but rather to abnormal ossification.

Evidence-based information on the ages of onset and completion of lacunar skull is lacking. All previous studies have reported that lacunar skull is evident at birth and disappears spontaneously within 4–6 months. We report for the first time that it persists to at least the age of 16 years and does not decrease, but rather increases, over time, becoming very pronounced with age in some children. If lacunar skull is attributable to a high intracranial pressure during the intrauterine period, as previously suggested, it is difficult to explain why it is still present (and becomes even more pronounced) in the absence of an increase in intracranial pressure at the age of 16 years. The current lack of better understanding of this condition is due to the lack of radiological data on the incidence of lacunar skull in healthy children.

Several studies have explored the incidences of intraspinal abnormalities in patients with idiopathic scoliosis. A meta-analysis reported a high overall frequency of neuro-axial abnormalities evident in MRI in patients with adolescent idiopathic scoliosis (8% in pooled analyses of >4,000 patients) [27]. Pereira et al. [28] reported that the incidence of intraspinal abnormalities was 5.6% in patients aged <10 years. Zhang et al. [29] studied patients <10 years of age and found that the prevalence of intraspinal neural axis abnormalities in those with presumed idiopathic scoliosis was 18.7%. Another study reported that this rate was 13.0% in children with infantile scoliosis [30]. Tully et al. [31] found that 14.7% of patients under 18 years of age with presumed idiopathic scoliosis but normal neurological examinations evidenced neuro-axial disease in full spinal MRI. Early-onset scoliosis is associated with a higher rate of neurological abnormalities. In the present study, as expected, lacunar skull and additional spinal and cranial anomalies were more common in infantile scoliosis patients <6 years of age. Therefore, we did not include these children.

We found that the incidence of neuro-axial abnormalities was 3.6% in children with lacunar skulls who underwent spinal MRI; 2 of 56 had isolated CMs-I. None required scoliosis surgery, and no increase in intracranial pressure was evident in cranial MRI. In addition, both the patients were neurologically normal. Given the high incidence of lacunar skull, if all patients had undergone spinal MRI, it is likely that stronger associations would have been evident between idiopathic scoliosis, lacunar skull, and CMs.

This study has several limitations. It was retrospective in nature, and we included both children and adolescents. Not all children with scoliosis and lacunar skull underwent full spinal MRI, only 20.9% of them did. The strengths of the study include the large sample size and the fact that all lacunar head images were saved. The medical files of all children with lacunar skulls were reexamined in terms of neurological disease. We believe

Fig. 6. An 11-year-old girl was diagnosed idiopathic scoliosis (right thoracic scoliosis, curve of 60°) and lacunar skull (clearly pronounced finger-shaped pits on the skull). MRI scan demonstrated a CM-I (tonsillar herniation into the foramen magnum, 6.08 mm).
that this study contributes to the literature on lacunar skull and emphasizes the need to examine the associations among lacunar skull, idiopathic scoliosis, intraspinal anomalies, and the need for MRI. However, there is a clear need for a prospective clinical study in which all children will be evaluated via X-ray, full spinal MRI, and neurologically.

**Conclusion**

Lacunar skull does not always indicate the presence of an intraspinal neural axis abnormality; it is a very common benign condition in patients with idiopathic scoliosis without any additional neurological pathology. In addition, it does not disappear over time, and in fact may become more obvious even up to the age of 16 years. Radiological data on the incidence of lacunar skull in healthy children are lacking. More studies are warranted to clarify whether the lacunar skull is a variant of normal skull development.

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**Statement of Ethics**

The Bezmialem Vakif University Local Research Ethics Committee approved this retrospective study and waived the requirement for written informed consent.

**Conflict of Interest Statement**

The author has indicated that there are no potential conflicts of interest to disclose.

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The author has indicated that there are no financial relationships relevant to this article to disclose.

**Author Contributions**

A.V. designed the study. A.V. collected data and wrote the manuscript.

**Data Availability Statement**

We interrogated the database of Bezmialem Vakif University Hospital (Istanbul).
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