MODERN DISEASES AND SAFETY ISSUES IN THE TREATMENT OF ONCOLOGICAL APLASIAS

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Annotatsiya

Bugungi kunda onkologik kasalliklar orasida aplaziya holatlari alohida ahamiyat kasb etmoqda. Aplaziya — bu hujayralar yoki to'qimalarning rivojlanmasligi yoki yo'qligi bilan kechadigan holat bo'lib, u bir qancha xavfli o'smalar bilan bevosita yoki bilvosita bog'liq bo'ladi. Ayniqsa, gematologik yo'nalishda uchraydigan aplastik anemiya, miya hujayralari aplaziyasi yoki limfoid to'qimalarning yetishmovchiligi kabi shakllar zamonaviy onkologik amaliyotda ko'plab muammolarni yuzaga keltirmoqda. Davolash jarayonida immunosupressiv terapiya, giyohvandlikka qarshi vositalar, transplantatsiya va genetik muvofiqlik muhim omillar bo'lib qolmoqda. Ushbu maqolada onkologik aplaziyalarning zamonaviy davolash usullari, ularning samaradorligi, shuningdek, mavjud muammolar va ilmiy izlanishlarga ehtiyoj tahlil qilinadi.

Kalit so'zlar: Onkologik aplaziya, aplastik anemiya, immunosupressiv terapiya, transplantatsiya, genetik moslik, dolzarb muammolar

Аннотация: В настоящее время случаи аплазии в онкологии приобретают особое значение. Аплазия — это состояние, при котором отсутствует развитие или формирование клеток и тканей, часто связанное с различными злокачественными новообразованиями. Особенно актуальны формы, встречающиеся в гематологии, такие как апластическая анемия, аплазия клеток костного мозга, дефицит лимфоидной ткани и другие. Современные подходы к лечению включают иммуносупрессивную терапию, противоопухолевые препараты, трансплантацию и генетическую совместимость. Однако данные методы не являются универсальными. В данной статье рассматриваются

современные методы терапии онкологических аплазий, их эффективность, а также существующие проблемы и необходимость дальнейших научных исследований.

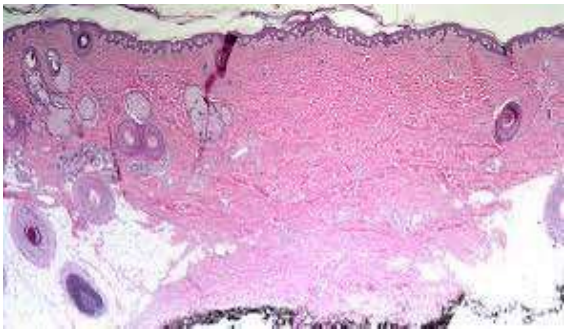
Ключевые слова: Онкологическая аплазия, апластическая анемия, иммуносупрессия, трансплантация, генетическая совместимость, актуальные проблемы.

Abstract: Nowadays, cases of aplasia in oncology are gaining increasing importance. Aplasia is a pathological condition characterized by the absence or underdevelopment of cells or tissues, and it is often directly or indirectly associated with malignant tumors. Particularly relevant are hematological forms such as aplastic anemia, bone marrow cell aplasia, and lymphoid tissue deficiency. Modern treatment approaches include immunosuppressive therapy, antitumor drugs, transplantation, and genetic matching. This article discusses current therapeutic strategies for oncological aplasias, evaluates their effectiveness, and highlights the urgent need for further scientific research.

Keywords: Oncological aplasia, aplastic anemia, immunosuppressive therapy, transplantation, genetic compatibility, pressing issues.

Introduction: Aplasia cutis congenita is a rare, congenital disorder. In its severe phenotype, it is potentially life threatening. Its management and the timing of surgery remain controversial because of the risks involved with both conservative and surgical approaches. Most literature is based on case reports and very small case series because of the rarity of the disorder. The authors present their experience treating newborns with aplasia cutis congenita and its progressive development. Aplasia Cutis Congenita (ACC), congenital absence of the skin, is an uncommon anomaly. It most commonly presents as a solitary defect of the scalp but may also involve the trunk and extremities. The lesions are non inflammatory, well demarcated and have variable extents, ranging birth. The cause is not clear but genetic factors, compromised vasculature to the skin, infection, teratogens, fetus papyraceous and trauma are all implicated. Truncal aplasia cutis congenita has been reported with biliary atresia, distal duodenal atresia, intestine infarction and multiple hepatic hematomas. Syndromes such as Adams Oliver syndrome, SCALP syndrome (nevus sebaceus, CNS malformations, aplasia cutis congenita, limbal dermoid, pigmented nevus). [1]

Histological details are available in very few reports. Histological features vary depending on the depth and duration of aplasia. Ulcers are seen at birth. After healing, the epidermis appears flattened with proliferation of fibroblasts within a connective tissue stroma. The total absence of the epidermal appendages remains a characteristic.


Aplasia cutis congenita histology

The diagnosis of ACC can typically be made solely from clinical examination. Many providers withhold performing a lesional biopsy given the patient's age demographics and typical scalp involvement. When a biopsy is necessary to aid in the diagnosis, proper workup, including imaging of the lesion with ultrasound (US) or MRI, is important to ensure there are no underlying malformations that can be damaged during the biopsy procedure. Histopathologic findings of non-healed lesions include an absent epidermis and/or dermis with a proliferation of blood vessels. A subtype known as membranous-type ACC will have a thin translucent membrane covering. Lesions that have already healed with a scar will have a thin or flattened epidermis, absent adnexal structures, and dense, dermal fibrosis noted on pathology.

A classification for ACC was proposed in 1986, which is still accepted today, and presented below. [2]

- Group 1: Scalp ACC without multiple anomalies
- Group 2: Scalp ACC with limb abnormalities
- Group 3: Scalp ACC with epidermal and organoid nevi
- Group 4: ACC overlying congenital malformations
- Group 5: ACC with associated fetus papyraceus or placental infarct
- Group 6: ACC with epidermolysis bullosa
- Group 7: ACC localized to extremities without blistering
- Group 8: ACC due to specific teratogens
- Group 9: ACC associated with malformation syndromes

ACC is a rare congenital condition with an incidence of approximately 1 to 3 out of 10,000 births.[3] There is no significant gender or cultural predilection that has been reported in the literature. [4] Lesions will typically be noticed at birth, although patients may not present to be evaluated for several months as lesions are often asymptomatic.

ACC can be associated with underlying morphologic abnormalities in approximately 37% of cases, according to Mesrati et al. including underlying bony defects, vascular anomalies, or neurologic malformations, so it is prudent for clinicians to evaluate the disease involvement with imaging. A midline vertex scalp lesion, hair collar sign, and vascular stains have all been shown to be strong indicators for cranial or central nervous system (CNS) involvement. Small, scalp lesions are less likely

associated with underlying defects and typically heal on their own within a couple of months; therefore, monitoring these lesions without further imaging is acceptable. For larger, ulcerative lesions, ultrasound provides a relatively inexpensive evaluation while not putting the child through a great deal of discomfort. If there is any concern for underlying defects on ultrasound, further workup with MRI is warranted. MRI is more sensitive and specific for identifying underlying lesions according to a 2017 retrospective multicenter study; however, it is more costly than ultrasound and typically requires the child to be sedated for the duration of the procedure, making this a poor choice for initial screening. If the lesion is purulent or surrounded by erythema, a lab workup including complete blood count, blood cultures, and wound cultures would be advised.

Although isolated ACC without an underlying defect can have a relatively benign course when complications occur, the risk of mortality dramatically increases. The estimated mortality rate ranges from 20% to 55% as a result of serious complications. The most common life-threatening complication of ACC is sagittal sinus bleeding, seen with lesions nearby on the scalp. Another potential complication of ACC includes secondary infection of the lesion. Patients are at an increased risk of cutaneous infections, given the fact that the skin's barrier against environmental microbes is absent or impaired. Severe infections can progress to meningitis if not treated appropriately. Prompt management of large scalp lesions, commonly with surgery, can help prevent these complications.

Aplasia Cutis Congenita: Clinical Types, Causes, and Management

No.	Type of ACC (Classification)	Appearance	Etiology	Associated Syndromes	Treatment Approach
1	Type I (only scalp)	Hairless scarred zone	Isolated (idiopathic)	None	Monitoring, no surgery needed
2	Type II (deep skin defect)	Full-thickness skin loss	Teratogens (drugs, infections)	Trisomy 13	Possible skin grafting
3	Type III (scalp and face)	Multiple lesion zones	Genetic – AD/AR patterns	Adams–Oliver syndrome	Multidisciplinary approach
4	Type IV (other body parts)	Neck, back, legs	Vascular disruption in skin	Limb-body wall complex	Evaluate for additional anomalies

5	Type V (twin-related)	Focal defect area	Twin gestation with circulatory issues	Fetus papyraceus	Esthetic and psychological support
6	Type VI (with EB)	Fragile, blistering skin	Autosomal recessive disorder	Epidermolysis bullosa	Wound care + protective dressings
7	Type VII (amniotic bands)	Band-like scar formation	Amniotic membrane rupture	Craniofacial defects	Surgical intervention required

Table 1.

Conclusion:

If imaging with ultrasound or MRI shows concern for an underlying abnormality, surgical intervention is usually necessary to prevent complications such as superior sagittal sinus hemorrhage, meningitis, thrombosis, among others. When isolated cases without underlying defects are present, the prognosis is quite good with simple wound care and close monitoring by pediatricians and/or dermatologists.[22] Although complications are rare, an understanding of ACC is important for providers to effectively work-up cases and to ensure prompt consultations are made when needed.

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